Comparing palliation strategies for single-ventricle anatomy with transposed great arteries and systemic outflow obstruction

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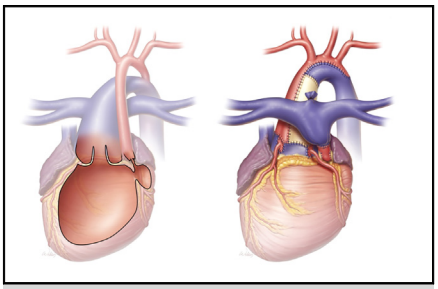
ABSTRACT

Objective: Patients with complex single-ventricle anatomy with transposed great arteries and systemic outflow obstruction (SV-TGA-SOO) undergo varied initial palliation with ultimate goal of Fontan circulation. We examine a longitudinal experience with multiple techniques, including the largest published cohort following palliative arterial switch operation (pASO), to describe outcomes and decision-making factors.

Methods: Neonates with SV-TGA-SOO who underwent initial surgical palliation from 1995 to 2022 at a single institution were retrospectively reviewed.

Results: In total, 71 neonates with SV-TGA-SOO underwent index surgical palliation at a median age of 7 days (interquartile range, 6-10) by pASO (n = 23), pulmonary artery band (PAB) with or without arch repair (n = 25), or modified Norwood with Damus–Kaye–Stansel aortopulmonary amalgamation (n = 23). Single-ventricle pathology included double-inlet left ventricle (n = 37, 52%), tricuspid atresia (n = 27, 38%), and others (n = 7, 10%). All mortalities (n = 5, 7%) occurred in the first interstage period after PAB (n = 3) and Norwood (n = 2). Subaortic obstruction in the PAB group was addressed by operative resection (n = 10 total, 7 at index operation) and/or delayed aortopulmonary amalgamation (n = 13, 52%). Two patients with pASO (9%) had early postoperative coronary complications, 1 requiring operative revision. Median follow-up for survivors was 10.4 years (interquartile range, 4.5-16.6 years). Comparing patients by their initial palliation type, notable significant differences included size of bulboventricular foramen, weight at initial operation, operation duration, postoperative length of stay, time to second-stage palliation, multiple pulmonary artery reinterventions, and left pulmonary artery interventions. There were no significant differences in overall survival, Fontan completion, reintervention-free survival in the first interstage period, pulmonary artery reintervention-free survival, long-term systemic valve competency, or ventricular dysfunction.

Conclusions: Excellent mid- to long-term outcomes are achievable following neonatal palliation for SV-TGA-SOO via pASO, PAB, and modified Norwood, with comparable survival and Fontan completion. Initial palliation strategy should be individualized to optimize anatomy and physiology for successful Fontan by ensuring an unobstructed subaortic pathway and accessible pulmonary arteries. pASO is a reasonable strategy to consider for these heterogeneous lesions. (JTCVS Techniques 2023;21:149-77)



Representative pre- and postoperative anatomy after palliative arterial switch operation.

CENTRAL MESSAGE

Fontan completion and survival are comparable for palliative arterial switch, pulmonary artery band, and modified Norwood for neonates with single-ventricle, transposition, and systemic obstruction.

PERSPECTIVE

Choice of initial surgical palliation for single-ventricle with transposition and systemic outflow obstruction is challenging and has long-reaching implications for successful Fontan. We review a diverse longitudinal experience, including the largest published series of the controversial palliative arterial switch operation, to highlight long-term outcomes and salient factors in decision-making.

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Abbreviations and Acronyms

BCPC = bidirectional cavopulmonary connection
BVF = bulboventricular foramen
BSA = body surface area
DKS = Damus–Kaye–Stansel
LPA = left pulmonary artery
MRI = magnetic resonance imaging
PA = pulmonary artery
PAB = pulmonary arterial banding
pASO = palliative arterial switch operation
PBF = pulmonary blood flow
RPA = right pulmonary artery
SPS = systemic-to-pulmonary artery shunt
SV-TGA-SOO = single-ventricle, transposed great arteries, and systemic outflow tract obstruction

For the rare subset of patients with single-ventricle, transposed great arteries, and systemic outflow obstruction (SV-TGA-SOO), successful Fontan palliation is dependent on controlling pulmonary blood flow (PBF), relieving obstruction in the systemic pathway, preserving ventricular and semilunar valve function, ensuring a stable rhythm, and allowing pulmonary arterial development. Difficulty in objectively assessing and predicting progression of subaortic obstruction in these heterogeneous lesions further complicates operative decision-making.

Typical palliation strategies in the neonatal period include pulmonary artery banding (PAB) with arch augmentation[1] or subaortic resection[2] as needed and modified Norwood procedure with Damus–Kaye–Stansel (DKS) aortopulmonary amalgamation and systemic-to-pulmonary shunt (SPS).[3] The palliative arterial switch operation (pASO) is an alternative approach that aligns the systemic ventricle with an unobstructed outflow tract, preserves antegrade pulmonary blood flow, and avoids left pulmonary artery (LPA) entrapment.[4] Critiques of this operation include its technical complexity and concern for coronary artery complications.

In this study, we describe nearly 3 decades of experience caring for neonates with SV-TGA-SOO at a high-volume center, including the largest published experience with pASO. By comparing patient characteristics and outcomes in this population, we aim to provide historical and contemporary context for early operative management in this challenging population.

METHODS

Study Design

This study was approved by the Baylor College of Medicine institutional review board (protocol H-38659, approved September 8, 2021 with waiver of informed consent). We queried the Texas Children's Hospital patient database for patients with diagnoses of single ventricle and transposition who underwent index surgical repair at age ≤30 days between July 1995 and July 2022. Medical records were retrospectively reviewed. Anatomic inclusion criteria were preoperative diagnoses consistent with single-ventricle cardiac configuration, transposition, and at least one level of systemic outflow obstruction (aortic arch hypoplasia, coarctation, or interruption; aortic valve hypoplasia or stenosis; or presence/substrate for subaortic obstruction due to hypoplastic subaortic chamber, restrictive/potentially restrictive bulboventricular foramen [BVF], or prominent subaortic conus). The latter criteria were deliberately broad, given lack of consensus on objective criteria for subaortic adequacy in the preoperative setting. Exclusion criteria were index operation at outside institution or biventricular repair.

A single cardiologist blinded to palliation strategy systematically reviewed preoperative echocardiograms, focusing on semilunar valves and subaortic region. Parameters were preferentially obtained by direct review of images and extrapolated from existing reports when images were unavailable (studies before 2010). Qualitative follow-up echocardiographic parameters (ventricular function, systemic valve insufficiency) were retrospectively obtained from existing reports.

Patients were primarily categorized and compared by their initial intervention (PAB, pASO, or Norwood), unless otherwise stated. Primary outcomes were Fontan completion and survival. Secondary outcomes were unplanned cardiothoracic interventions in the first interstage period, pulmonary artery (PA) interventions, postoperative coronary artery pathology, interventions for subaortic obstruction, transplant, ventricular function, and systemic valve regurgitation.

Strategy Selection

Choice of initial palliation technique was individualized to each patient's anatomy and hemodynamics, informed by preoperative multidisciplinary discussion, intraoperative transesophageal echocardiography, and direct anatomic inspection. Arrangement of the great vessels, coronary pattern, presence/degree of subaortic obstruction, and semilunar valve function were major determinants of the decision. The aggregate balance of risks, benefits, and feasibility of each option was interpreted subjectively by the performing surgeon, often finalizing the decision intraoperatively. Further details regarding the decision-making process are subsequently discussed, and current practices are summarized in Figure 1.

Operative Technique: Palliative Arterial Switch

Following median sternotomy, institution of cardiopulmonary bypass, and cardioplegic arrest, the ascending aorta was transected several millimeters distal to the aortic valve commissures. If necessary to obtain adequate coronary buttons, native aortic valve commissures were taken down and reapproximated. Neopulmonary sinuses were reconstructed with autologous pericardial patches. Coronary transfer was typically accomplished via medially based trapdoor incisions. Two of the three surgeons adjusted their approach during the study period to reconstruct the neoaorta before performing coronary transfer. A Lecompte maneuver was performed in all patients. Arch reconstruction was performed when indicated, typically by arch advancement with or without patch augmentation. After weaning from bypass, relative PBF was assessed and restricted (PAB) or augmented (SPS) to maintain saturations between 75% and 85% and distal PA pressures approximately half systemic. Sternal closure was delayed for hemodynamic instability or anticipated need for early PBF adjustment.

Operative Technique: Pulmonary Artery Band

PAB were placed distally on the main PA to minimize distortion of native pulmonary valve and sized by Trusler's rule, tailored based on

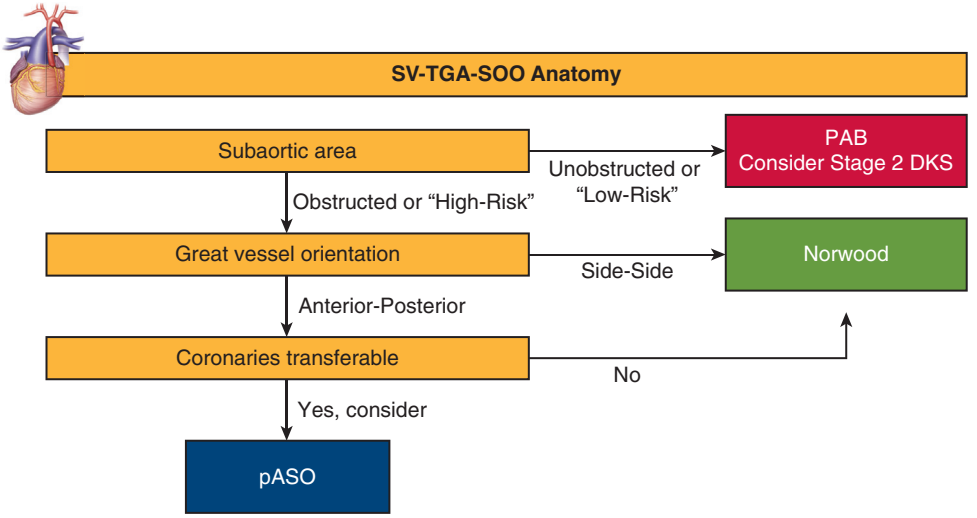


FIGURE 1. Algorithm summarizing main anatomic factors driving initial palliation strategy selection at Texas Children's Hospital in the current era, although additional patient factors and surgeon preferences continue to guide the final decision. PAB with subaortic resection is generally avoided as an initial strategy in current practice, given risk for conduction system injury and recurrent obstruction. *SV-TGA-SOO*, Single-ventricle, transposed great arteries, and systemic outflow tract obstruction; *PAB*, pulmonary artery banding; *pASO*, palliative arterial switch operation.

systemic oxygen saturation and echocardiographic gradient across the band. Concurrent repair of arch obstruction and other cardiac lesions were performed when indicated. Strategies for dealing with preoperative presence or threat of subaortic obstruction shifted during the study period, with subaortic resection (BVF enlargement, septal myectomy, or resection of accessory valve tissue) to enlarge the subaortic area at index PAB not performed since 2015.

Operative Technique: Modified Norwood

End-to-side or side-to-side ("double-barrel") DKS aortopulmonary amalgamation was performed, depending on great vessel arrangement, to create the neoaortic root. Patch augmentation was performed as needed to avoid semilunar valve distortion and address arch obstruction. The distal PA bifurcation was either closed primarily or patch augmented. SPS were modified Blalock–Thomas–Taussig shunts, except one case with central shunt. The Lecompte maneuver was performed in one case.

Statistical Analysis

Continuous variables were summarized as medians and interquartile ranges and compared with Kruskal–Wallis or Wilcoxon rank-sum tests. Categorical variables were described as absolute and relative frequencies within the group of interest and compared using Fisher exact test. Differences in Kaplan–Meier survival curves for time-to-event outcomes were evaluated by log-rank testing. Post-hoc pairwise analyses were performed for statistically significant multi-group comparisons, without adjustment. Analyses were conducted with STATA, version 17.0 (StataCorp LLC).

RESULTS

Preoperative Characteristics

Selected preoperative characteristics are summarized in Tables 1 and E1. Demographics, fundamental cardiac diagnosis, great vessel looping and orientation, and arch pathology were similarly distributed between groups. Primary single-ventricle lesions were double-inlet left ventricle (n = 37, 52%), tricuspid atresia (n = 27, 38%), and other arrangements (n = 7, 10%) involving double-outlet right ventricle or other complex congenitally-corrected transposition.

On preoperative echocardiography (Tables 1, E2, and E3, and Figures E1-E3), there was a nearly statistically significant trend ($P = .052$) toward greater prevalence of pulmonary valve pathology, defined as greater than mild insufficiency and/or leaflet thickening, in the PAB group. Prevalence of subaortic conus and BVF was similar between groups. Although a similar proportion of patients had potentially restrictive BVF by size criteria (BVF/body surface area [BSA] < 2 cm^2/m^2),[5] minimum BVF diameter (Figure E1), absolute BVF area (Figure E2), and indexed BVF area to BSA (Figure E3) significantly differed, largest in the PAB group, particularly for those without initial subaortic resection.

Index Operation and Interstage Period

Operative data are summarized by cohort in Tables 2 and E4, and Figure E4. Although operative age was similar, operative weight was heavier for patients with Norwood compared with PAB. Cardiopulmonary bypass and crossclamp times were longest in pASO, followed by Norwood then PAB. Delayed sternal closure was more common after pASO and Norwood compared with PAB. Two patients (1 pASO, 1 Norwood) had extracorporeal membrane oxygenation cannulation at initial palliation for ventricular dysfunction. Both required 4 days of support and were alive at follow-up.

Postoperative events in the first interstage period between initial palliation and bidirectional cavopulmonary connection (BCPC) are detailed in Tables E5 and E6. Postoperative length of hospital stay was shortest for PAB but similar for pASO and Norwood (Table 2). Freedom from any

TABLE 1. Preoperative characteristics

Variable	Overall (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB (n = 25)	*P* value
Primary cardiac diagnosis					.57
DILV	37 (52%)	12 (52%)	10 (43%)	15 (60%)	
TA	27 (38%)	10 (43%)	9 (39%)	8 (32%)	
Other (DORV, cc-TGA)	7 (10%)	1 (4%)	4 (17%)	2 (8%)	
Great vessel looping					.40
D-TGA	43 (61%)	12 (52%)	13 (57%)	18 (72%)	
L-TGA	28 (39%)	11 (48%)	10 (43%)	7 (28%)	
Great vessel orientation					.92
Anterior-posterior	59 (83%)	20 (87%)	19 (83%)	20 (80%)	
Side-side	12 (17%)	3 (13%)	4 (17%)	5 (20%)	
Interrupted aortic arch	12 (17%)	5 (22%)	5 (22%)	2 (8%)	.49
Coarctation	46 (65%)	14 (61%)	17 (74%)	15 (60%)	.89
Pulmonary valve pathology*	12 (17%)	2 (9%)	2 (9%)	8 (32%)	.052
Subjective subaortic obstruction	15 (21%)	4 (17%)	8 (35%)	3 (12%)	.16
Subaortic conus	51 (72%)	19 (83%)	15 (65%)	17 (68%)	.25
BVF	64 (90%)	21 (91%)	20 (87%)	23 (92%)	.89
BVF/BSA, cm^2/m^2	0.87 (0.54-1.36) n = 40	0.66 (0.37-1.14) n = 14	0.69 (0.53-1.11) n = 13	1.37 (1.07-1.96) n = 13	**.002**
Potentially restrictive BVF (BVF/BSA <2)	35 (49%)	12 (50%)	13 (57%)	10 (40%)	.58

N (% total) or median (interquartile) with n = nonmissing data when applicable. Multigroup comparison *P* value reported, with values < .05 bolded. Subaortic and pulmonary valve parameters reported based on preoperative echocardiography. Additional demographic data (Table E1), echocardiographic data (Tables E1-E3; Figures E1-E3) and pairwise comparisons (Table E3) are reported in the Appendix. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *DILV*, double-inlet left ventricle; *TA*, tricuspid atresia; *DORV*, double-outlet left ventricle; *cc-TGA*, congenitally corrected transposition of the great arteries; *D-TGA*, dextro transposition of the great arteries; *L-TGA*, levo transposition of the great arteries; *BVF*, bulboventricular foramen; *BSA*, body surface area. *Pulmonary valve pathology defined as greater than mild insufficiency or leaflet thickening.

unplanned intervention (related to cardiovascular pathology or postsurgical complications) or death before BCPC was statistically similar between groups ($P = .08$, Figure 2, *A*), although tended to be superior for PAB.

Interventions to adjust relative pulmonary and systemic blood flow are summarized in Figure 3. Of the 14 patients with pASO with PAB placed at index procedure, 3 required PAB adjustment postoperatively (2 before chest closure), and 2 required SPS for increasing BVF restriction and PAB. A total of 4 patients with pASO had SPS placed at a median of 40 days (range, 3-77 days) postoperatively.

Mortality

There were 5 total mortalities (3 PAB, 2 Norwood), all occurring in the first interstage period at a median of 68 days postoperatively (range, 22-245 days). Mortality rates were statistically similar between cohorts (Table 2). One patient with PAB developed increasing BVF restriction and pulmonary hypertension and underwent conversion to DKS with SPS 4 months postoperatively (BCPC was attempted but aborted due to profound hypoxia). Although PA pressures improved, multifactorial chronic respiratory failure and ventricular dysfunction persisted until their death 8 months following initial palliation. Another patient developed complete heart block following PAB and subaortic resection requiring pacemaker placement; they died from a sudden respiratory arrest 2 months postoperatively. At 2-month follow-up for the third patient with PAB, significant ventricular dysfunction with wall motion abnormalities, ST changes, and elevated troponin were discovered. Workup revealed patent coronaries but multiple venous thrombi. They developed complete heart block and suffered cardiac arrest, followed by acute ischemic stroke with hemorrhagic conversion and persistent poor ventricular function. Ultimately, they were withdrawn from transplant listing and died 8 months postoperatively. A patient who initially presented in extremis with severe ventricular dysfunction before Norwood died less than 1 month postoperatively. The second patient with a Norwood had intrauterine drug exposure and an accessory anomalous coronary artery branch from the PA; their postoperative course was complicated by recurrent severe sepsis, renal failure, and hemodynamic lability despite good cardiac function, and they died 1 month postoperatively.

Fate of the Coronary Arteries

Clinical details of patients with any coronary artery pathology following initial palliation (4 pASO, 3 Norwood) are summarized in (Table E7). Two patients had coronary complications related to the pASO: 1 patient required revision of right coronary anastomosis 38 days following pASO for symptomatic proximal stenosis. During the initial operation, another patient with abnormal coronary anatomy

TABLE 2. Single-ventricle palliation

Variable	pASO (n = 23)	Norwood (n = 23)	PAB (n = 25)	*P* value
Index palliation				
Operative age, d	7 (5-9)	7 (6-9)	8 (7-13)	.19
Operative weight, kg	3.3 (2.9-3.6)	3.5 (3.3-3.9)	3.1 (2.9-3.5)	**.03** *.4** *.053*† ***.02***‡
CPB time, if used, min	272 (221-292)	198 (173-211)	118.5 (95-153)	**.0001** *<**.0001***†‡
Postoperative LOS, d	23 (19-49)	26 (15-38)	19 (10-25)	**.015** ***.01**** *.50*† ***.02***‡
Interstage mortality (% total)	0 (0%)	2 (9%)	3 (12%)	.36
Any unplanned interstage intervention (% total)	10 (43%)	8 (35%)	5 (20%)	.23
BCPC (% total)	23 (100%)	21 (91%)	22 (88%)	.36
BCPC age, mo	4.3 (3.7-5.9)	4.5 (3.9-5.6)	7.4 (6.6-9.3)	**.0002** ***.0001**** *.31*† ***.0005***‡
BCPC PA plasty (% BCPCs)	16 (70%)	12 (57%)	11 (50%)	.45
Fontan (% total)	19 (83%)	18 (78%)	18 (72%)	.73
Fontan age, y	4 (3.5-4.6)	4.2 (3.6-4.7)	4.4 (3.8-5.0)	.79
Fontan PA plasty (% Fontans)	12 (63%)	6 (33%)	4 (22%)	**.04** ***.02**** *.10*† *.71*‡
Fontan co-operation (non-PA; % Fontans)	6 (33%)	2 (11%)	10 (56%)	**.02** *.19** *.23*† ***.01***‡

Number of patients (% total or at risk, as noted) or median (interquartile range). *P* value for difference in proportion or median among the 3 cohorts, with post-hoc pairwise comparisons included for significant differences (italics). Bold denotes raw *P* value <.05. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *CPB*, cardiopulmonary bypass; *LOS*, length of stay; *BCPC*, bidirectional cavopulmonary connection; *PA*, pulmonary artery. *pASO vs PAB. †pASO vs Norwood. ‡PAB vs Norwood.

underwent revision of left coronary anastomosis and extracorporeal membrane oxygenation cannulation due to concern for coronary insufficiency. Postoperative catheterization showed preserved distal flow with possible proximal left circumflex dissection (medically managed with therapeutic anticoagulation without intervention). On latest assessment, all 7 patients with coronary pathology were alive with patent coronaries and 6 had normal ventricular function.

In total, 21 patients with pASO (91%) had postoperative angiographic assessment of their coronaries (catheterization, computed tomography, or magnetic resonance imaging [MRI]), most recently performed at a median of 3.4 years (1.3-10.7 years) postoperatively; all studies showed patent coronaries.

Subaortic Obstruction After PAB

Management of subaortic obstruction in the PAB group is summarized in Figure 4. Ten patients (40%) had at least 1 subaortic resection during the study period, of whom 4 had multiple resections. Resections were performed at initial palliation (7/25, 28%), BCPC (4/23, 17%—3 without previous resection), and Fontan (3/18, 17%—all with previous resections). In total, 13 (52%) of patients with PAB underwent subsequent DKS, 5 due to persistent/recurrent subaortic obstruction after resection. DKS was generally performed at BCPC (n = 11), although 1 was performed post-Fontan after multiple previous resections and 1 was the interstage conversion to DKS/SPS (deceased).

Seven patients with PAB (28%) had no subaortic interventions during the study: 1 patient died in the first interstage period secondary to ventricular dysfunction of unclear etiology, 1 is planned to undergo DKS at upcoming Fontan, 2 are monitored for stable mild obstruction, and 3 were unobstructed at follow-up.

PA Interventions

In total, 48 patients (68%) had at least one PA intervention during the study period (operative patch augmentation,

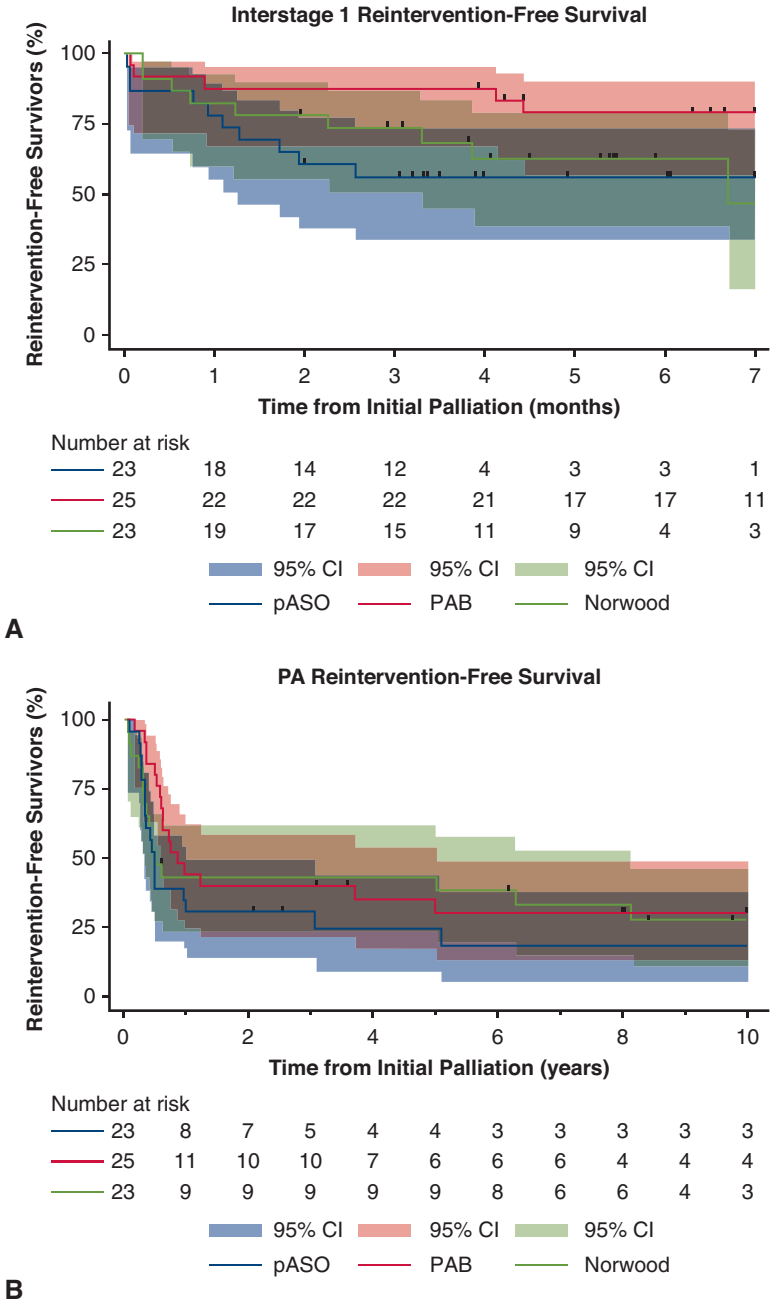


FIGURE 2. Kaplan–Meier survival curves with shaded 95% confidence intervals (*CI*), truncated at time points when approximately 10% of the starting populations remained at risk. A, Reintervention-free survival in the first interstage period, defined as time from initial palliation to either death or first unplanned intervention for cardiovascular pathology or postoperative complications, censored at second stage palliation with BCPC. There were no statistically significant differences between cohorts ($P = .08$). B, Pulmonary-artery reintervention-free survival, defined as time from initial palliation to death or first pulmonary artery angioplasty following initial palliation (catheter-based or operative), censored at last follow-up. There were no significant differences between cohorts ($P = .34$). *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *PA*, pulmonary artery.

catheter-based balloon angioplasty, or stent placement), and 21 had multiple PA interventions (Tables 3 and E8). Median time to first intervention was 5.9 months following initial palliation (4-11.2 months). Freedom from any PA angioplasty or death was similar for all cohorts ($P = .34$, Figure 2, *B*). Compared with PAB, a greater proportion of patients with pASO had LPA intervention(s) and multiple PA interventions.

PA augmentation as part of planned staged palliation was performed at 39 of 66 BCPC (59%) and 22 of 55 Fontan

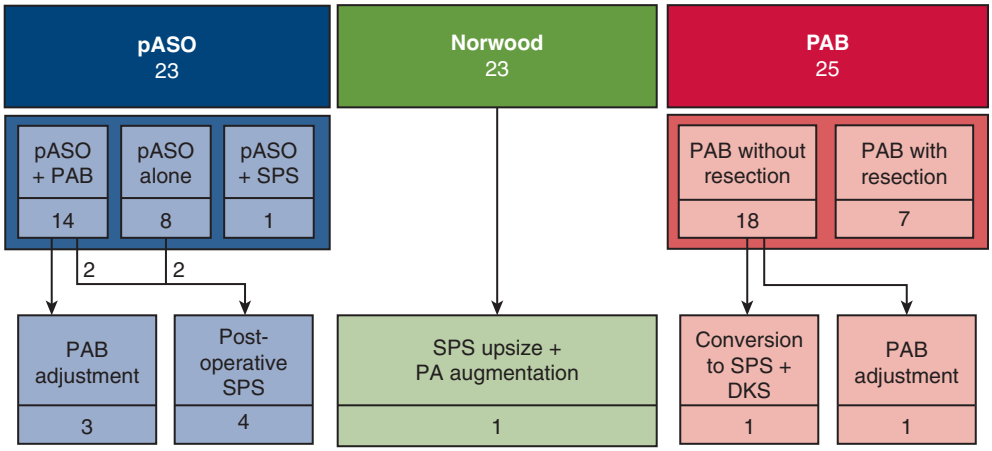


FIGURE 3. Schematic flow diagram showing patients' initial operative palliation (*top two rows of boxes*) and postoperative interventions for adjustment of relative pulmonary and systemic blood flow (*bottom row*). *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *SPS*, systemic-to-pulmonary artery shunt; *DKS*, Damus–Kaye–Stansel.

(40%). Rates of any PA augmentation and rates of LPA augmentation at BCPC were statistically similar between groups, although they tended to be more common for pASO (Table E8, and Figure E5). Rates of PA augmentation at Fontan differed ($P = .04$), significantly greater for pASO than PAB ($P = .02$).

On pre-BCPC imaging (catheterization, computed tomography, or MRI), minimum right pulmonary artery (RPA) and LPA diameters (absolute and indexed to BSA) were similar by cohort (Table E8, and Figure E5). Those who underwent RPA patch augmentation at BCPC had significantly smaller RPA diameters than those who did not ($P = .03$, Tables E8 and E9, and Figure E5), indicating a correlation between RPA size and the decision to intervene it at BCPC. However, LPA diameters were similar whether or not patients had BCPC LPA augmentation.

Comparing patients with pASO with anterior-posterior great vessels and Norwood patients with anterior-posterior great vessels, no significant differences in pre-BCPC PA dimensions or rates of PA interventions (although the latter were higher for pASO, Table E10).

A total of 36 patients had a DKS during the study period (23 Norwood, 13 PAB), of whom 21 (58%) had any subsequent PA intervention and 8 (22%) had LPA intervention. Among the 35 patients without DKS, 25 (71%) had any PA intervention and 21 (60%) had an LPA intervention. Comparing those with and without a DKS, there were similar rates of PA intervention ($P = .32$), but fewer LPA interventions in those with a DKS ($P = .002$).

Continued Palliation and Follow-up

All 66 surviving patients completed BCPC (Table 2). Age at BCPC was older for the PAB cohort compared with pASO and Norwood. 55 patients (83% of survivors) completed Fontan, with similar rates by cohort. Of the 11 patients awaiting Fontan, 3 had pre-Fontan cardiac catheterization (2/4 pASO, 1/4 PAB, and 0/3 Norwood), all with appropriate hemodynamics (Tables E5 and E6). The remainder were awaiting catheterization or following at other institutions. More patients with PAB than Norwood had other concomitant procedures at Fontan (Table 2).

There was an association between arrhythmia requiring pacemaker or antiarrhythmic medication at most recent follow-up and having had subaortic resection or BVF enlargement ($P = .03$): 6/11 patients (55%) with previous resection had arrhythmias compared with 12 of 60 patients (20%) without.

Median follow-up time for survivors was 10.4 years (4.5-16.6 years). Seven patients were lost to follow-up with no documented visits in the last 5 years (Table E12). Nine patients (13%) required any coarctation reintervention (balloon angioplasty or operation) following index palliation, with a nonsignificant trend toward greater rates after Norwood (Table 3). Ventricular function and systemic valve competency were uniformly well-preserved between groups (Tables 3, E12, and E13, and Figures E6 and E7). At most recent echocardiography, 85% of survivors had normal ventricular function and 95% survivors had at most qualitatively mild systemic valve insufficiency. Three patients had moderate neoaortic valve insufficiency after pASO, all stable without indication for intervention at recent follow-up. One patient required cardiac transplant due to ventricular dysfunction and failing Fontan physiology (9 years post-Fontan, 12 years after pASO).

COMMENT

For complex single-ventricle patients with malposed great arteries and systemic outflow obstruction, selecting

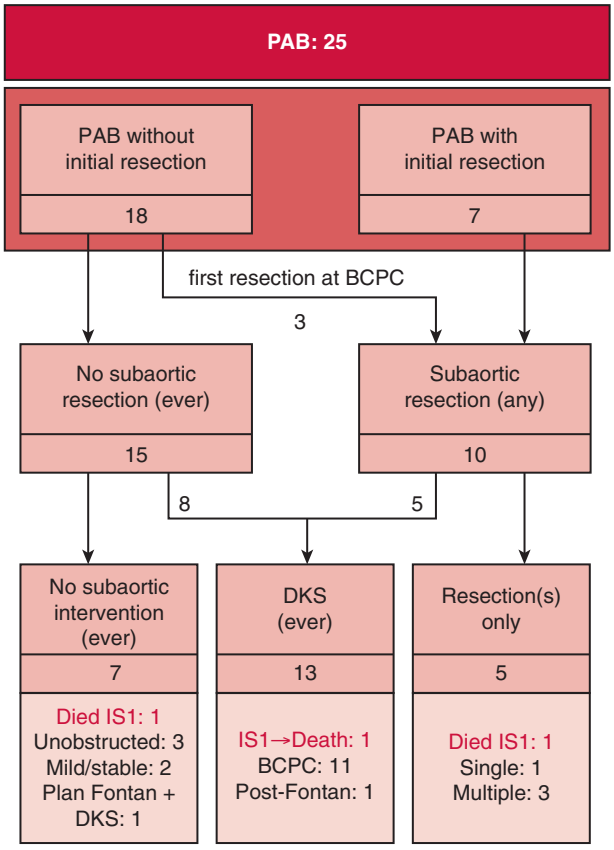


FIGURE 4. Diagram summarizing interventions for subaortic obstruction in the PAB cohort during the study period. Practices shifted away from performing subaortic resection or BVF enlargement, given the associated risks of conduction system injury and recurrent obstruction. In current practice, we prefer either Norwood or pASO as initial palliation if substantial subaortic obstruction is present or felt to be likely to develop in the short term, rather than initial PAB with resection. For those with lower risk or unobstructed subaortic regions, PAB may be performed initially with delayed DKS at time of BCPC if subaortic obstruction develops or there is ongoing concern for future obstruction. *PAB*, Pulmonary artery banding; *BCPC*, bidirectional cavopulmonary connection; *DKS*, Damus–Kaye–Stansel.

the optimal strategy for initial palliation is challenging and carries far-reaching implications for Fontan success. With such a heterogeneous population, no single strategy can address the needs of every patient: a given anatomic configuration may be composed of certain factors advantageous to one strategy but disadvantageous to others. Based on our long-term experience with this population, we aim to summarize the advantages, drawbacks, and ideal patient for each management strategy (Figure 5). By comparing preoperative characteristics of patients selected for each strategy, we highlight the salient objective and subjective features that impact early operative decision-making. Although outcome comparisons between these groups are fraught with inherent limitations, they are meant to contextualize our center's continued use of pASO and inform those considering including it in current practice.

Challenges and Changes in Approach to Subaortic Obstruction

Substrates for subaortic obstruction in SV-TGA-SOO include hypoplastic subaortic outflow chamber, BVF, and subaortic conus. Multiple sites of obstruction and/or accompanying shunts limit the use of gradient or flow acceleration across the subaortic region or BVF to quantify the degree of preoperative obstruction. Size of the interventricular communication indexed to BSA or aortic valve diameter has been proposed as a potential predictor of current or future restriction,[5-7] but this does not take into account other obstructive configurations. Indeed, prevalence of subaortic conus and potentially restrictive BVF by size criteria were similar between groups, but BVF size was significantly larger in the PAB group, particularly those who did not have initial subaortic resection. Rather than a single parameter, an aggregate judgment regarding the risk for subaortic obstruction is made based on objective size parameters coupled with subjective anatomic assessment. In addition to traditional echocardiography and direct operative inspection, advanced imaging modalities including 3-dimensional echocardiography and MRI may be a useful adjunct for delineating subaortic anatomy.

For patients with existing or high-risk for early postoperative subaortic obstruction, the index procedure must provide adequate systemic outflow in order to prevent the catastrophic effects of increased ventricular pressure load (hypertrophy) and low cardiac output state (coronary insufficiency, systemic malperfusion).[6-9] In current practice, we feel this is best accomplished with modified Norwood or pASO. Historically, subaortic resection with PAB was considered a reasonable strategy,[2,10] although it has fallen out of favor due to risk for conduction system injury and recurrent obstruction.[11] Of the 10 patients with PAB who underwent subaortic resection in our study, 4 required multiple resection procedures (1 of whom underwent DKS post-Fontan for recurrent obstruction despite several previous resections) and 5 had arrhythmia requiring pacemaker or antiarrhythmic medication at last follow-up. In addition, these procedures may be technically difficult in those with tricuspid atresia, necessitating ventriculotomy to perform the resection, which adds risk to the procedure.

Even if the subaortic area is adequate at initial palliation, the trajectory remains difficult to predict, with time to development of obstruction ranging from 10 days to 15 years postoperatively.[12] In our cohort of 25 patients with PAB with SV-TGA-SOO, only 3 had no intervention to relieve the threat or presence of subaortic obstruction and remained unobstructed at last follow-up. Proposed mechanisms for BVF restriction after PAB are altered ventricular geometry following acute volume unloading and hypertrophy due to increased afterload,[6] effects that may be compounded by pre-existing arch obstruction.[9] Delayed DKS (typically at BCPC)[10,13] is an effective solution, but

TABLE 3. Follow-up

Variable	Overall (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB: All (n = 25)	*P* value
Follow-up time (y, survivors)	10.4 (4.5-16.6)	7.9 (4.0-16.2)	9.8 (4.5-16.6)	13.0 (6.0-17.8)	.50
Any PA intervention* (% total)	48 (68%)	18 (78%)	15 (65%)	15 (60%)	.40
Any LPA intervention (% total)	39 (55%)	17 (74%)	13 (57%)	9 (36%)	**.04** ***.01***† *.35*‡ *.25*§
Multiple PA interventions (% total)	21 (30%)	11 (48%)	7 (30%)	3 (12%)	**.03** ***.01***† *.37*‡ *.16*§
Arrhythmia‖					
% total	18 (25%)	3 (13%)	4 (17%)	11 (44%)	**.03**
% survivors	16 (24%)	3 (13%)	4 (19%)	9 (41%)	.09
Any coarctation reintervention (% total)	9 (13%)	2 (9%)	5 (22%)	2 (8%)	.37
Ventricular dysfunction (mild or greater, % survivors)	10 (15%)	4 (17%)	3 (14%)	3 (14%)	1.0
Normal-"low-normal"	56 (85%)	19 (83%)	18 (86%)	19 (86%)	.81
Mildly depressed	8 (12%)	4 (17%)	2 (10%)	2 (9%)	
Moderately depressed	2 (3%)	0 (0%)	1 (5%)	1 (5%)	
Systemic valve insufficiency¶ (mild or greater, % survivors)	35 (53%)	13 (57%)	13 (62%)	9 (41%)	.37
None-trivial	31 (47%)	10 (43%)	8 (38%)	13 (59%)	.15
Mild	32 (48%)	10 (43%)	13 (62%)	9 (41%)	
Moderate	3 (5%)	3 (13%)	0 (0%)	0 (0%)	

Number of patients (% total or at risk, as noted) or median (interquartile range). *P* value for difference in proportion or median among the 3 cohorts, with post-hoc pairwise comparisons in italics. Bold denotes statistical significance with raw *P* value <.05. Additional details for pulmonary artery imaging and interventions available in the supplement (Tables E8-E11 and Figure E5). Additional follow-up details and echocardiographic data found in Tables E12 and E13, Figures E6 and E7. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *PA*, pulmonary artery; *LPA*, left pulmonary artery; *DKS*, Damus–Kaye–Stansel. *PA interventions defined as operative PA augmentation or catheter-based balloon angioplasty or stent placement. †pASO vs PAB. ‡pASO vs Norwood. §PAB vs Norwood. ‖Arrhythmia requiring antiarrhythmic medication or pacemaker placement at last follow-up. ¶Neoaortic valve insufficiency for those with DKS or pASO, native aortic valve for those with PAB and no DKS.

vigilant monitoring with reliable follow-up is critical to ensure intervention is performed before subaortic obstruction progresses to ventricular hypertrophy, heart failure, or coronary insufficiency. Individual surgeons may differ in their subjective judgment assessment and tolerance of risk for future subaortic obstruction. Some may prefer to eliminate any possibility of future obstruction by performing DKS at second stage palliation more routinely after initial PAB, whereas others may be comfortable continuing to observe whether there is no subaortic obstruction at time of second-stage palliation. Similar biases also affect the choice of initial palliation, balancing the relative simplicity of initial PAB with the risks associated with unpredictable subaortic obstruction.

Pulmonary Artery Banding

PAB with arch augmentation when necessary is the least-complex operative technique, boasting shorter bypass times and length of stay in our study. In addition to the aforementioned complexity of managing evolving subaortic obstruction with resection or subsequent DKS, there is potential for encroachment of the band on the branch PAs or native pulmonary valve.

Patients with unobstructed or low-risk subaortic areas are the best candidates for initial PAB. Other factors that make Norwood or pASO more technically challenging or suboptimal may favor PAB, including pulmonary valve pathology, coronary anatomy that would be compromised by DKS or not amenable to switch, or inability to tolerate long operations or shunted physiology.

Modified Norwood

The modified Norwood procedure has been advocated by multiple centers[11,14-16] as a solution that effectively addresses any existing or potential subaortic obstruction at the index operation. Progress is evident with reduction in neoaortic regurgitation[17] with the adoption of the Lamberti ("double-barrel") modification of DKS.[14] Era-related advances in experience and critical care are credited with improving the once substantial interstage morbidity and mortality associated with this technique.[8,18] Regardless, early commitment to completely shunt-dependent pulmonary blood flow carries substantial complication risk and often requires prolonged and complex postoperative care. In our study, one patient required shunt upsizing and PA augmentation for insufficient pulmonary blood flow and

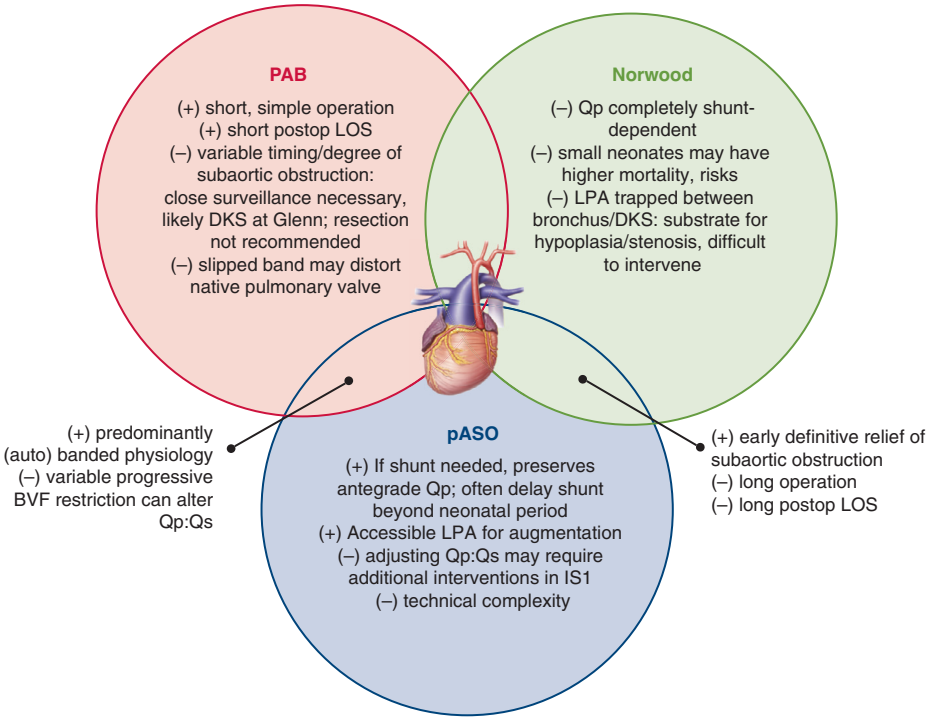


FIGURE 5. Venn diagram summarizing the major advantages (+) and disadvantages (-) of 3 palliation techniques for SV-TGA-SOO. *SV-TGA-SOO*, Single-ventricle, transposed great arteries, and systemic outflow tract obstruction; *PAB*, pulmonary artery banding; *LOS*, length of stay; *DKS*, Damus–Kaye–Stansel; *LPA*, left pulmonary artery; *pASO*, palliative arterial switch operation; *BVF*, bulboventricular foramen; *Qp-Qs*, ratio of pulmonary to systemic blood flow.

another was found to have shunt thrombosis at BCPC. Patients in the Norwood cohort were heavier at initial operation, reflecting our preference to avoid SPS physiology in smaller neonates.

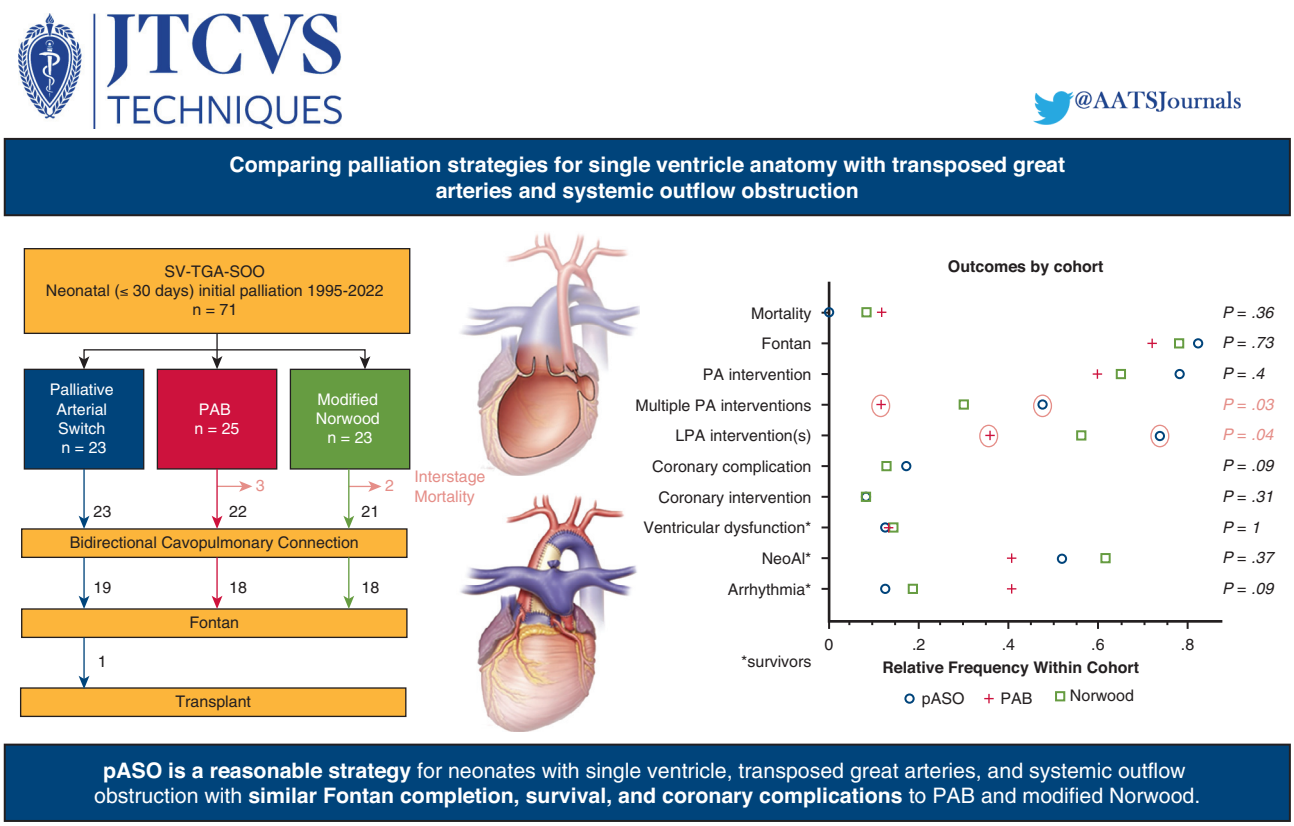
An additional concern regarding early SPS and DKS is potential compromise to PA growth[3,19,20] and accessibility. Particularly in the setting of anterior-posteriorly oriented great vessels, the LPA is trapped between the neoaorta and left mainstem bronchus. This configuration provides substrate for compression and growth limitation and complicates options for intervention, should significant stenosis develop. Operative LPA augmentation may require DKS transection and reconstruction, and it cannot address space constraints. Catheter-based intervention is possible, although balloon angioplasty often provides incomplete or temporary relief and stents are at risk for erosion into neoaorta, bronchial compression, or stent fracture due to external compression.

Overall, we feel the modified Norwood approach is best suited to patients with existing or greater risk of subaortic obstruction (as DKS is preferable to resection), side-by-side great arteries (preserves more space for LPA development), and coronary anatomy unfavorable to translocation. Drawbacks to this technique include commitment to shunt-dependent PBF and its associated risks, as well as entrapment of the LPA (which has potential to limit Fontan candidacy and require extensive and higher risk intervention, should stenosis develop).

Palliative Arterial Switch

The pASO was first proposed as an alternative strategy for patients with SV-TGA-SOO in the 1980s.[21] It has subsequently been successfully applied as initial palliation in the neonate and infant population[4,22-24] with successful promotion to Fontan.[4,23-25] Critiques of this approach include longer operative times, intraoperative complexity, and concern for coronary artery complications. Proposed benefits include alignment of the dominant semilunar valve with the systemic circulation, eliminating the need for BVF enlargement, postponement or avoidance of shunt-dependent physiology, and preserved LPA accessibility.

Direct alignment of the functional single ventricle with the systemic circulation creates an unobstructed, laminar outflow tract and physiologically "auto-banded" circulation by converting subaortic obstruction to subpulmonary obstruction. Of our 23 patients with pASO, 14 (60%) had initial PAB and 8 were "auto-banded" (6 remained well-balanced without requiring PAB or SPS). Shunt physiology was avoided or postponed past the vulnerable immediate postoperative setting for most patients (of 5 patients with pASO with SPS, only 1 was placed at index operation). Even when SPS is required, it avoids completely shunt-dependent circulation by preserving antegrade PBF, which may allow for improved symmetric PA growth. Achieving balanced systemic and pulmonary blood flow may be challenging, and the potential need for postoperative adjustment



LPA: left pulmonary artery; neo-AI: neo-aortic valve insufficiency; PA: pulmonary artery; PAB: pulmonary artery band; pASO: palliative arterial switch operation; SV-TGA-SOO: single ventricle with transposed great arteries and systemic outflow obstruction

FIGURE 6. Outcomes including mortality, Fontan completion, coronary complications and interventions, ventricular function, and neoaortic valve function were similar between the surgical palliation cohorts. A greater proportion of patients with pASO had LPA intervention(s) and multiple PA reinterventions compared with PAB, possibly reflecting the easy accessibility for pulmonary artery augmentation following pASO. These results imply that pASO is a reasonable strategy for initial palliation in neonates with SV-TGA-SOO with comparable outcomes to PAB and modified Norwood. *SV-TGA-SOO*, Functional single ventricle, transposed great arteries, and systemic outflow tract obstruction; *PA*, pulmonary artery; *PAB*, pulmonary artery banding; *LPA*, left pulmonary artery; *pASO*, palliative arterial switch operation.

of PAB or placement of SPS should be anticipated. Delayed chest closure allows opportunities to intervene once pulmonary congestion has improved. However, the now-subpulmonary obstruction may continue to evolve beyond the immediate postoperative period.

Our previously published favorable short-term experience with the pASO[4] has been sustained at longer-term follow-up with 100% survival. Overall, patients with coronary anatomy not amenable to translocation, morphologically abnormal native pulmonary valve, or side-by-side great vessels may be better served by other palliation strategies. However, for patients with subaortic obstruction (or at significant risk) and anterior-posterior great vessels, pASO can safely and effectively palliate these complex patients with superior accessibility of the branch PAs for ease of augmentation at staged operations. Drawbacks to the pASO include the potential for coronary complications, unpredictable PBF, and a lengthy, complex operation. Although our findings confirm that indeed, cardiopulmonary bypass and aortic crossclamp times are longest in the pASO group, postoperative length of stay was similar to the Norwood group. Furthermore, coronary complications were rare and did not impact long-term coronary patency or survival. Among these complex single ventricle patients, coronary artery anomalies are common, and may require intervention or monitoring regardless of palliation strategy.

PA Interventions: Accessibility or Pathology?

Given the selection bias inherent in the study, our data do not definitively answer the debate regarding the true impact of LPA entrapment by DKS on PA growth. Future imaging studies in expanded cohorts of patients may shed further light on PA growth following DKS versus Lecompte. LPA diameters were not significantly different between pASO and Norwood prior to BCPC, even comparing the subgroup of patients with anterior-posterior great vessels.

Of the 23 patients with pASO, 18 (78%) underwent at least 1 PA angioplasty and 11 had multiple. In total, 17

had at least 1 operative augmentation, including 16 at BCPS and 12 at Fontan. A total of 17 patients had at least one intervention that included the LPA. Significant differences between the cohorts in rates of LPA intervention, multiple PA interventions, and PA angioplasty at Fontan reached pairwise significance only between PAB and pASO, with similar rates between pASO and Norwood (though pASO was observed to have the highest rates in these categories). Patients with DKS had similar any-side PA interventions but fewer subsequent LPA interventions compared with those without DKS.

The underlying explanations for these differences are likely multifactorial. Our institution has a liberal policy toward PA augmentation (68% of patients had any PA intervention and PA augmentation was performed at 59% of BCPC and 40% of Fontan operations). The accessibility of the PAs after Lecompte may confer a lower threshold to intervene at planned palliation and the LPA may be compressed as it drapes over the neoaorta. Conversely, there may be a greater threshold to intervene for those with impaired accessibility due to DKS, requiring relatively more severe stenosis or hypoplasia to justify the intervention. Our pre-BCPC imaging data showed that although RPA size correlated with RPA intervention at BCPC, the same was not true for the LPA, indicating that factors other than vessel size likely influence the decision to intervene upon the LPA.

Weighing the Options

Overall, meaningful mid- to long-term outcomes for all 3 palliation strategies were comparable (Figure 6). Mortality rates were similar, with no late deaths. All surviving patients progressed to BCPC and either completed or are awaiting Fontan. On pre-Fontan evaluation, all groups had effective protection of their pulmonary vasculature with low pulmonary artery pressures, low transpulmonary gradient, and low pulmonary arteriolar resistance as well as preserved ventricular function with normal end diastolic pressures. Only 1 patient required late post-Fontan transplant.

Our outcomes demonstrate that pASO facilitates safe, effective palliation to a Fontan circulation, making it a valuable addition to the surgeon's armamentarium in approaching these complex patients. Although not appropriate for every patient, we believe the pASO is a reasonable option to consider for patients with subaortic obstruction, anterior-posterior great vessels, and transferrable coronary arteries. In these challenging situations, no single technique has proven superior, and patient selection criteria for each palliation strategy remain ill-defined. We therefore recommend individualized assessment of each patient's anatomy and hemodynamics pre- and intraoperatively in order to select an approach to create a favorable circulation for successful Fontan.

Limitations

This study is subject to the limitations of a retrospective single-center design as well as the heterogeneity of the complex cardiac lesions. Data were obtained from the medical chart, operative notes, echocardiographic reports, and catheterization reports which vary in reporting standardization and completeness. The limited number of neonatal patients with SV-TGA-SOO palliated at our institution during this time period could bias the results toward finding no difference between groups when there actually is a difference. Low number of events such as death and transplantation precluded multivariable regression analysis. The long duration of the study in order to characterize the historical and contemporary experience does add complexity with changing practices over time. In addition, there is significant inherent selection bias among the groups, with subtle and often competing anatomic factors influencing subjective decision-making. Results and comparisons should be interpreted within the context of these limitations as a descriptive longitudinal experience rather than a rigorously matched comparison.

CONCLUSIONS

The palliative arterial switch is a valuable addition to the surgical armamentarium for neonates with single ventricle, transposition, and systemic outflow obstruction. Although not appropriate for every patient, pASO is a reasonable option to consider for safe and effective palliation of patients, especially those with subaortic obstruction, anterior-posterior great vessels, and transferrable coronary arteries. Excellent survival and Fontan completion were comparable to modified Norwood and PAB, and pASO confers PA accessibility for ease of augmentation without substantial risk of coronary artery complications.

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: palliative arterial switch, complex single ventricle, single-ventricle management, double-inlet left ventricle, tricuspid atresia, transposition

APPENDIX E1: COMPARING PALLIATION STRATEGIES FOR SINGLE VENTRICLE, TRANSPOSED GREAT ARTERIES, AND SYSTEMIC OUTFLOW OBSTRUCTION

This supplement provides full, detailed parameters and additional subgroup analysis to complement that included in the main article to allow more summative and descriptive figures as part of the main text.

Note: *P* values are raw multigroup comparisons unless otherwise stated, results reported as n/N at-risk (%) and median (IQR) unless noted otherwise, with n = number with non-missing and/or applicable data. Statistical significance (raw *P* value < .05) bolded.

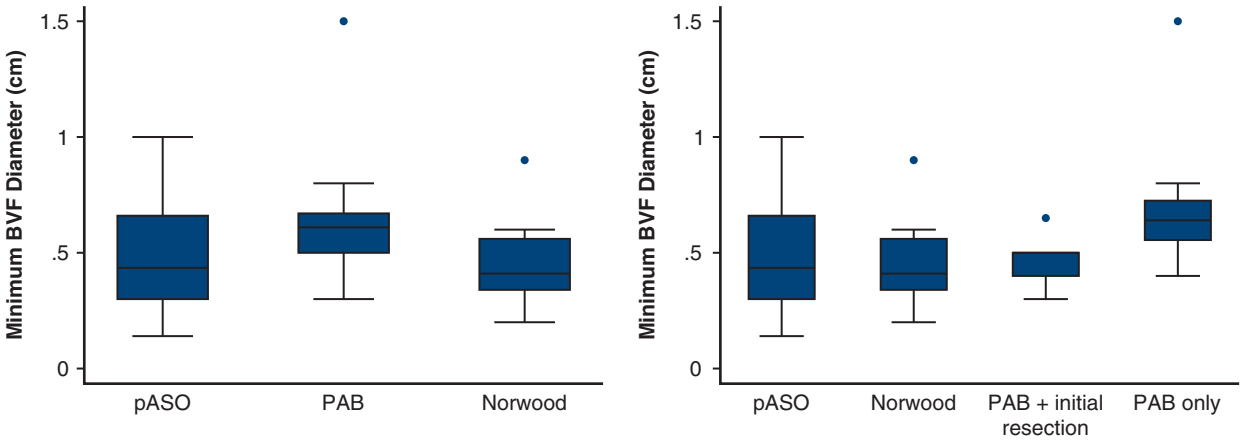

FIGURE E1. Minimum BVF diameter by initial palliation. *BVF*, Bulboventricular foramen; *pASO*, palliative arterial switch operation; *PAB*, pulmonary artery banding.

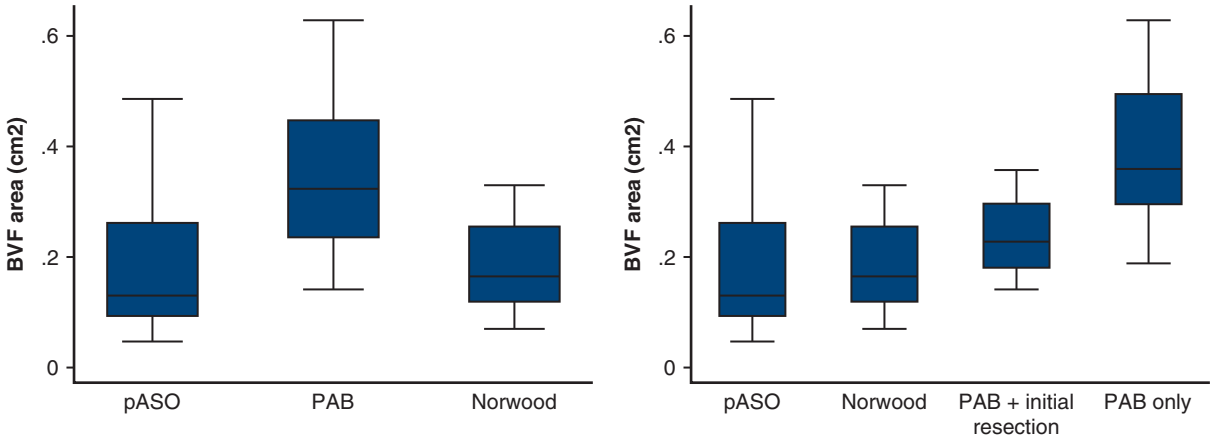

FIGURE E2. BVF area by initial palliation. *BVF*, Bulboventricular foramen; *pASO*, palliative arterial switch operation; *PAB*, pulmonary artery banding.

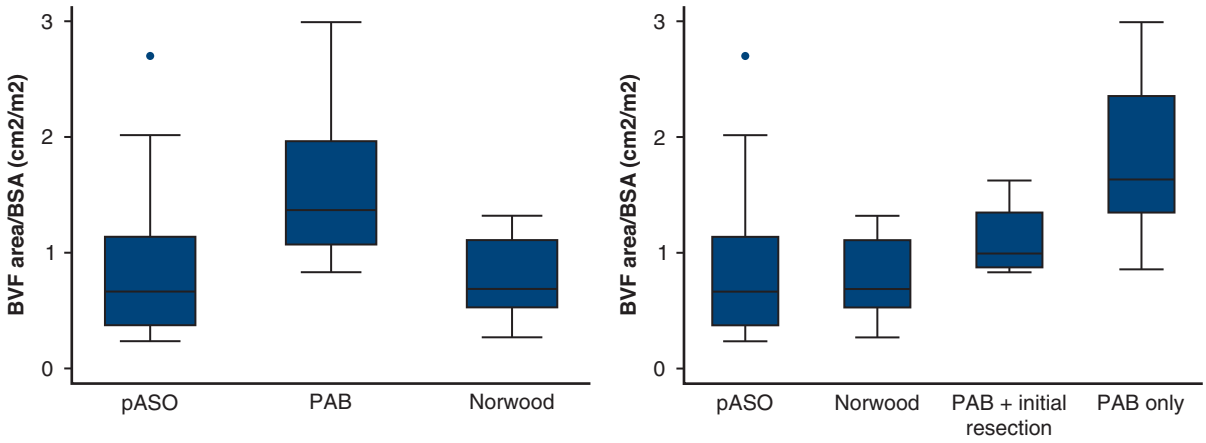

FIGURE E3. BVF/BSA by initial palliation. *BVF*, Bulboventricular foramen; *BSA*, body surface area; *pASO*, palliative arterial switch operation; *PAB*, pulmonary artery banding.

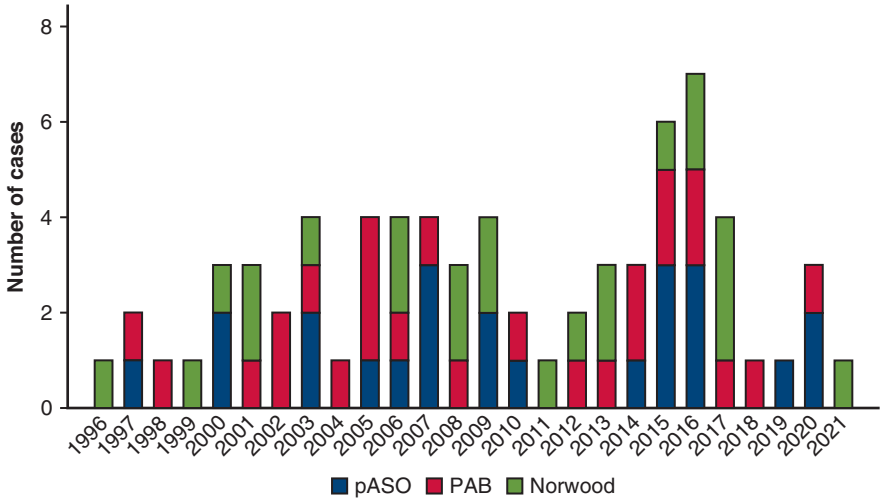

FIGURE E4. Operation types over time. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding.

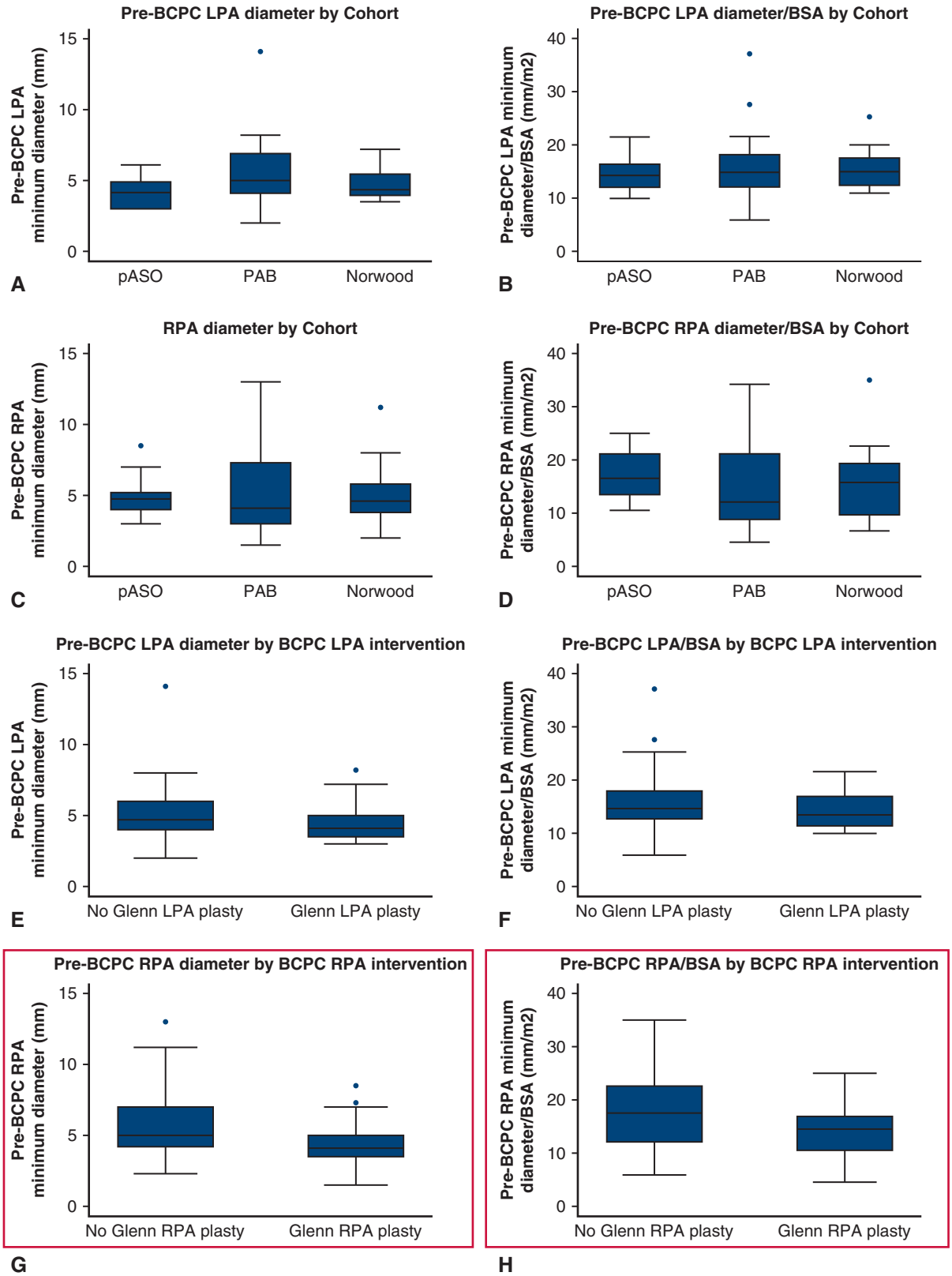

FIGURE E5. Pre-BCPC PA imaging by cohort and BCPC intervention (see Tables E8 and E9). Box plots demonstrating absolute (*left*) and indexed to BSA (*right*) minimum PA diameters on pre-BCPC imaging by cohort (A-D) and by whether or not PA augmentation on that side was performed at BCPC (E-H). For the LPA (A-B, E-F) and RPA (C-D, G-H). Plots outlined in *red* had statistically significant differences between the groups shown. *BCPC*, Bidirectional cavopulmonary connection; *LPA*, left pulmonary artery; *BSA*, body surface area; *pASO*, palliative arterial switch operation; *PAB*, pulmonary artery banding; *RPA*, right pulmonary artery.

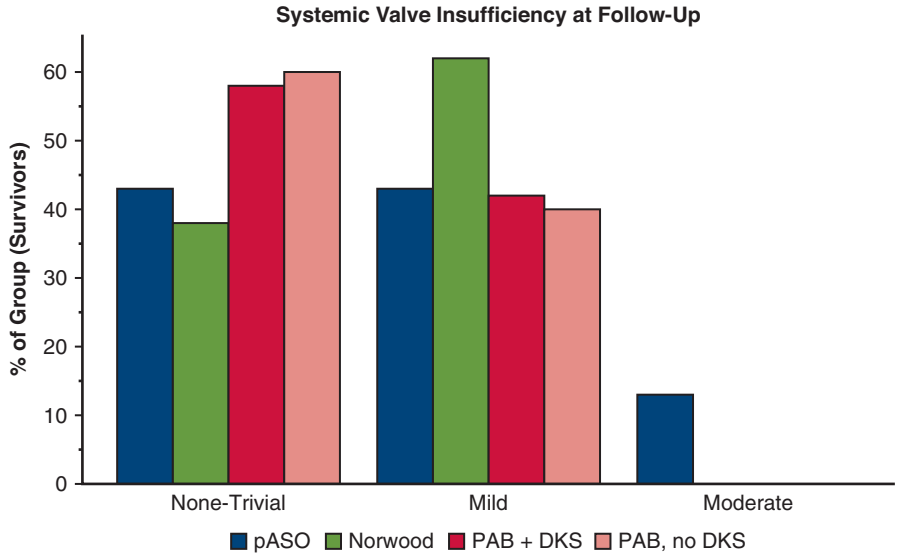

FIGURE E6. Systemic valve insufficiency by PAB subgroups (with vs without DKS). *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *DKS*, Damus–Kaye–Stansel.

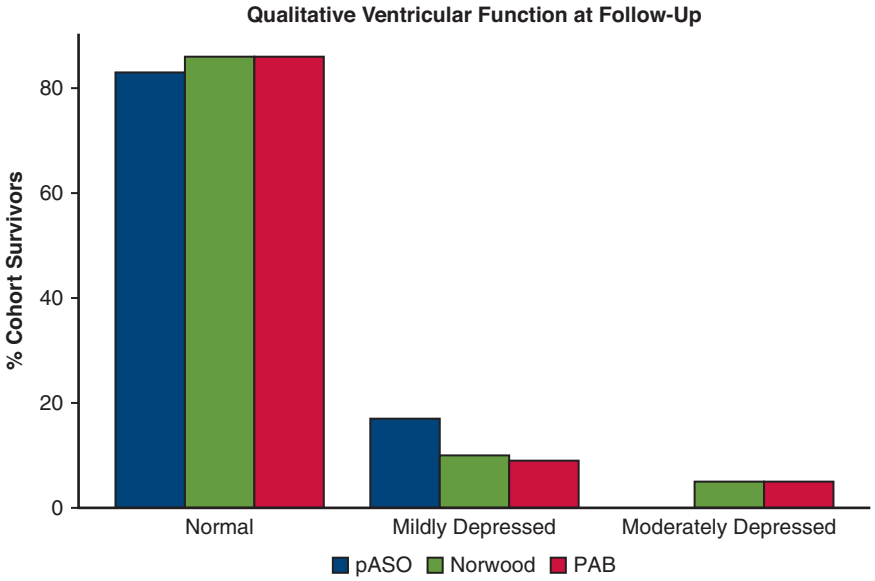

FIGURE E7. Ventricular dysfunction at follow-up by cohort. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding.

TABLE E1. Patient demographics and anatomy

Variable	Total (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB (n = 25)	*P* value
Male	62 (87%)	20 (87%)	20 (87%)	22 (88%)	1.00
Premature (<37 wk)	5 (7%)	2 (9%)	2 (9%)	1 (4%)	.73
Birth weight, kg	3.4 (3.0-3.7) n = 63	3.4 (2.8-3.6) n = 23	3.6 (3.3-3.8) n = 18	3.3 (2.9-3.5) n = 22	.06
Genetic syndrome	17 (24%)	5 (22%)	7 (30%)	5 (20%)	.69
Chromosomal abnormality	12 (17%)	7 (30%)	2 (9%)	3 (12%)	.24
Extracardiac anomaly	10 (14%)	3 (13%)	2 (9%)	5 (20%)	.20
Primary cardiac diagnosis					
DILV	37 (52%)	12 (52%)	10 (43%)	15 (60%)	.57
TA	27 (38%)	10 (43%)	9 (39%)	8 (32%)	
Other (DORV, cc-TGA)	7 (10%)	1 (4%)	4 (17%)	2 (8%)	
TGA looping					.40
D-TGA	43 (61%)	12 (52%)	13 (57%)	18 (72%)	
L-TGA	28 (39%)	11 (48%)	10 (43%)	7 (28%)	
Great vessel relationship					.92
Anterior-posterior	59 (83%)	20 (87%)	19 (83%)	20 (80%)	
Side-side	12 (17%)	3 (13%)	4 (17%)	5 (20%)	
Interrupted aortic arch	12 (17%)	5 (22%)	5 (22%)	2 (8%)	.49
Coarctation	46 (65%)	14 (61%)	17 (74%)	15 (60%)	.89
Yacoub coronary type					.50
A	37 (52%)	14 (61%)	8 (35%)	15 (60%)	
B	3 (4%)	1 (4%)	1 (4%)	1 (4%)	
C	3 (4%)	2 (9%)	1 (4%)	0 (0%)	
D	2 (3%)	1 (4%)	0 (0%)	1 (4%)	
E	1 (1%)	0 (0%)	0 (0%)	1 (4%)	
Other	22 (31%)	5 (22%)	10 (43%)	7 (28%)	

This is an expanded version of Table 1 in the article, including demographic characteristics and coronary patterns. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *DILV*, double-inlet left ventricle; *TA*, tricuspid atresia; *DORV*, double-outlet left ventricle; *cc-TGA*, congenitally corrected transposition of the great arteries; *D-TGA*, dextro transposition of the great arteries; *L-TGA*, levo transposition of the great arteries.

TABLE E2. Complete preoperative echocardiographic data with PAB subgroups

Variable	Total (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB: All (n = 25)	*P* value: 3 groups	PAB + initial resection (n = 7)	PAB without initial resection (n = 18)	*P* value: 4 groups
Echo images available	30/71 (42%)	10/23 (43%)	9/23 (39%)	11/25 (44%)	.96	2/7 (29%)	9/18 (50%)	.79
Semilunar valves								
AoV hypoplasia (z score < –2.5)	12/71 (17%)	5/23 (22%)	3/23 (13%)	4/25 (16%)	.79	0 (0%)	4 (22%)	.60
AoV/PV diameter, %	65.7 (57.3-78.4) n = 64	65.9 (56.3-80) n = 23	65.2 (61.1-71.3) n = 17	70.7 (56.0-81.7) n = 24	.71	77.4 (61.3-80.9) n = 7	70.4 (53.1-84) n = 17	.79
AoV annulus diameter, cm	0.61 (0.52-0.72) n = 65	0.56 (0.49-0.7) n = 23	0.6 (0.55-0.64) n = 18	0.66 (0.6-0.75) n = 24	.11	0.7 (0.63-0.82) n = 7	0.62 (0.57-0.73) n = 17	.07
AoV z score	−1.43 (−2.14- −0.065) n = 64	−1.5 (−2.43- −0.17) n = 23	−1.61 (−2.18- −1.16) n = 18	−0.57 (−1.69 - −0.33) n = 23	.17	0.33 (−0.67-0.88) n = 7	−1.24 (−2.26- −0.06) n = 16	.06
PV annulus diameter, cm	0.93 (0.83-1.06) n = 69	0.9 (0.81-1.06) n = 23	0.9 (0.87-1.01) n = 21	0.94 (0.82-1.07) n = 25	.84	1.03 (0.94-1.08) n = 7	0.88 (0.78-1.07) n = 18	.39
PV z score	0.625 (−0.02-1.48) n = 68	0.42 (0.08-1.5) n = 23	0.6 (0.01-0.93) n = 21	1.04 (−0.095-1.67) n = 24	.78	1.43 (1.01-2.37) n = 7	0.11 (−0.2-1.15) n = 17	.11
PV pathology*	12/71 (17%)	2/23 (9%)	2/23 (9%)	8/25 (32%)	.052	0/7 (0%)	8/18 (44%)	**.009**
Subaortic region								
Subjective subaortic obstruction†	15/71 (21%)	4/23 (17%)	8/23 (35%)	3/25 (12%)	.16	2/7 (29%)	1/18 (6%)	.11
Subaortic conus present	51/71 (72%)	19/23 (83%)	15/23 (65%)	17/25 (68%)	.25	6/7 (86%)	11/18 (61%)	.64
BVF present	64/71 (90%)	21/23 (91%)	20/23 (87%)	23/25 (92%)	.89	6/7 (86%)	17/18 (94%)	.84
Any measurement available	49/64 (77%)	18/21 (86%)	14/20 (70%)	17/23 (74%)	.50	5/7 (71%)	12/18 (67%)	.64
Two-plane measurements available	40/64 (63%)	14/21 (67%)	13/20 (65%)	13/23 (57%)	.81	4/7 (57%)	9/18 (50%)	.84
Minimum BVF diameter, cm	0.49 (0.35-0.65) n = 49	0.44 (0.3-0.66) n = 18	0.41 (0.34-0.56) n = 14	0.61 (0.5-0.67) n = 17	**.03**	0.5 (0.4-0.5) n = 5	0.64 (0.56-0.73) n = 12	**.02**
BVF area, cm^2‡	0.20 (0.11-0.31) n = 40	0.13 (0.09-0.26) n = 14	0.16 (0.12-0.26) n = 13	0.32 (0.24-0.45) n = 13	**.003**	0.23 (0.18-0.30) n = 4	0.36 (0.30-0.49) n = 9	**.003**
BVF/BSA, cm^2/m^2	0.87 (0.54-1.36) n = 40	0.66 (0.37-1.14) n = 14	0.69 (0.53-1.11) n = 13	1.37 (1.07-1.96) n = 13	**.002**	0.99 (0.87-1.35) n = 4	1.63 (1.35-2.35) n = 9	**.003**
Potentially restrictive BVF (BVF/BSA <2)§	35/71 (49%)	12/23 (50%)	13/23 (57%)	10/25 (40%)	.58	4/7 (57%)	6/18 (33%)	.12

P values all for multigroup comparisons: 3-group comparisons included pASO, Norwood, and all PAB; 4-group comparisons included pASO, Norwood, PAB with initial subaortic resection, and PAB without initial subaortic resection. Bold denotes raw *P* value < .05. Pairwise comparisons for significant differences reported in Table E3. BVF measurements visualized across groups in Figures E1-E3. Data obtained by single reviewer blinded to palliation status by review of images preferentially, and of existing imaging report when no images were available (no studies performed before 2010 were available for review). See the subheading "Study Design" in the "Methods." *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *AoV*, aortic valve; *PV*, pulmonary valve; *BVF*, bulboventricular foramen; *BSA*, body surface area. *Pulmonary valve pathology defined as greater than mild insufficiency and/or greater than mild leaflet thickening. †Subjective subaortic obstruction determined by reviewer judgment of subaortic area appearance or mentioned subaortic obstruction in imaging report. ‡BVF area calculated from measurements in 2 orthogonal planes. §BVF/BSA ratio < 2 used as a cutoff based on previous publication (see Matitiau and colleagues[5]) proposing this as a threshold for high risk of obstruction.

TABLE E3. Pairwise comparisons for echo data

Groups compared	PV pathology	Min BVF diameter	BVF area	BVF/BSA
pASO vs Norwood	1.0	.39	.36	.49
pASO vs PAB	–	**.01***	**.0008***	**.001***
Norwood vs PAB	–	**.01***	**.003***	**.001***
pASO vs PAB + resection	1.0	.42	.13	.12
pASO vs PAB without resection	**.01**	**.003***	**.0003***	**.0005***
Norwood vs PAB + resection	1.0	.35	.19	.12
Norwood vs PAB without resection	**.01**	**.002***	**.001***	**.0006***
PAB with vs without resection	.057	**.04**	.08	.11

Raw/unadjusted *P* values reported for preoperative echocardiographic parameters (Table E2) with multi-group *P* values <.05. Note, the additional groups considered in pairwise comparisons can increase type I error rate: *P* values < unadjusted type I error rate .05 are highlighted in bold. *PV*, Pulmonary valve; *BVF*, bulboventricular foramen; *BSA*, body surface area; *pASO*, palliative arterial switch operation; *PAB*, pulmonary artery banding. *Persistent significance against an adjusted type I error rate of 0.008 (4-group comparisons) or 0.02 (3-group comparisons).

TABLE E4. Initial palliation operative details

Variable	pASO (n = 23)	Norwood (n = 23)	PAB (n = 25)	*P* value
Patient size				
Operative age, d	7 (5-9)	7 (6-9)	8 (7-13)	.19
Operative weight, kg	3.3 (2.9-3.6)	3.5 (3.3-3.9)	3.1 (2.9-3.5)	**.03** *.4** *.053*† ***.02***‡
Operative times				
CPB used	23 (100%)	23 (100%)	22 (88%)	.10
CPB time, if used (mins)	272 (221-292)	198 (173-211)	118.5 (95-153)	**.0001** ***<.0001****†‡
Crossclamp used	23 (100%)	23% (100%)	22 (88%)	.10
Crossclamp time, if used, min	179 (138-208)	103 (83-128)	53.5 (41-67)	**.0001** ***<.0001****†‡
ACP used	20 (87%)	18 (78%)	17 (71%)	.39
ACP time, if used, min	49 (36-70)	78 (59-85)	21 (16-27)	**.0001** ***<.001****‡ *.10*†
DHCA used	16 (70%)	22 (96%)	19 (76%)	.06
DHCA time, if used	8 (6-14.5)	8.5 (6-15)	10 (5-24)	.90
Procedure details				
Arch repair	22 (96%)	23 (100%)	21 (84%)	.12
Arch patch	16 (70%)	17 (74%)	0 (0%)	**<.0001** ***<.0001****‡ *1.0*†
Atrial septectomy	18 (78%)	20 (87%)	18 (72%)	.51
Subaortic resection	0 (0%)	1 (4%)	7 (28%)	**.004** ***.01**** *1*† *.05*‡
Delayed sternal closure	15 (65%)	10 (43%)	2 (8%)	**<.0001** ***<.0001**** *.24*† ***.01***‡
ECMO cannulation	1 (4%)	1 (4%)	0 (0%)	.54
Era				
Operation year	2009 (2005-2016)	2009 (2003-2016)	2008 (2004-2015)	.84
Era				.80
1996-2008	10 (43%)	10 (44%)	13 (52%)	
2009-2021	13 (57%)	13 (57%)	12 (48%)	

Bold denotes raw *P* value < .05. Post-hoc pairwise comparisons included for significant differences (italics). *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *CPB*, cardiopulmonary bypass; *ACP*, antegrade regional cerebral perfusion time (minutes); *DHCA*, deep hypothermic circulatory arrest; *ECMO*, extracorporeal membrane oxygenation support. *pASO vs PAB. †pASO vs Norwood. ‡PAB vs Norwood.

TABLE E5. Single-ventricle palliation operations

Variable	Overall (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB (n = 25)	*P* value
Interstage 1					
Postoperative hospital length of stay, d	21 (15-35)	23 (19-49)	26 (15-38)	19 (10-25)	**.02** ***.01**** *.50*† ***.02***‡
Early readmission	20	9 (39%)	8 (35%)	3 (12%)	.07
Mortality	5	0 (0%)	2 (11%)	3 (14%)	.36
Unplanned IS1 intervention (any)§	23	10 (43%)	8 (35%)	5 (20%)	.23
Wound (any)	4	3 (13%)	1 (4%)	0 (0%)	.12
Re-coarctation (any)	7	2 (9%)	4 (17%)	1 (4%)	.29
PA (any)	4	1 (4%)	2 (9%)	1 (4%)	.84
Pacemaker (any)	2	0 (0%)	1 (4%)	1 (4%)	1.0
Multiple unplanned interventions§	4	3 (13%)	0 (0%)	1 (4%)	.21
Stage 2 palliation					
BCPC (100% survivors completed BCPC)	66/71 (93%)	23/23 (100%)	21/23 (91%)	22/25 (88%)	.36
Time from index palliation to BCPC, mo	5.3 (3.9-7.0)	4.0 (3.3-5.6)	4.3 (3.8-5.4)	7.0 (6.3-9.1)	**.0002** ***.0001**** *.69*† ***.0002***‡
Age at BCPC, mo	5.5 (4.1-7.4)	4.3 (3.7-5.9)	4.5 (3.9-5.6)	7.4 (6.6-9.3)	**.0002** ***.0001**** *.31*† ***.0005***‡
Weight at BCPC, kg	6.9 (5.9-8.1)	6.1 (5.5-7.4)	6 (5.7-8.1)	7.7 (6.8-8.4)	**.003** ***.0006**** *.32*† ***.004***‡
BCPC performed during same hospitalization as index palliation	5/66 (8%)	3/23 (13%)	2/21 (10%)	0/22 (0%)	.27
Pulsatile BCPC	5/66 (8%)	4/23 (17%)	0 (0%)	1/22 (5%)	.11
BCPC co-operation (any)	54/66 (82%)	17/23 (74%)	16/21 (76%)	21/22 (95%)	.11
Postoperative LOS, d	7 (6-9)	7 (6-8)	7 (6-9)	7 (6-9)	.77
Pre-Fontan hemodynamics					
Pre-Fontan cath	57/66 (86%)	18/23 (78%)	18/21 (86%)	21/22 (95%)	
PA pressure, mm Hg	11 (9-12)	11 (9.5-12)	11 (7.5-14)	11 (7.5-14)	.81
TPG, mm Hg	4 (3-5)	4 (3-5)	4 (3-5)	4 (3-5)	.85
PVR (Woods Units·m^2)	1.2 (0.9-1.7)	1.23 (0.9-1.65)	1.25 (1.01-1.75)	1.25 (1.01-1.75)	.42
EDP, mm Hg	8 (6-10)	8 (6-10)	8 (6-9)	8 (6-9)	.82
Qp:Qs	0.6 (0.5-0.7)	0.6 (0.5-0.7)	0.6 (0.5-0.7)	0.6 (0.5-0.7)	.69
Fontan					
Fontan completed (survivors)	55/66 (83%)	19/23 (83%)	18/21 (86%)	18/25 (82%)	1.0
Age at Fontan, y	4.2 (3.5-5.0)	4 (3.5-4.6)	4.2 (3.6-4.7)	4.4 (3.8-5.0)	.79
Fontan cooperation (any)	34/55 (62%)	14/19 (74%)	7/18 (39%)	13/18 (72%)	.06
Fontan cooperation (non-PA)	18/55 (33%)	6/19 (33%)	2/18 (11%)	10/18 (56%)	**.02**
Fenestrated Fontan	8/55 (15%)	4/19 (21%)	3/18 (17%)	1/18 (6%)	.50
Postoperative LOS, d	10 (8-12)	10 (8-12)	9 (8-11)	13 (8-18)	.10

Bold denotes raw *P* value < .05. Post-hoc pairwise comparisons included for significant differences (italics). *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *IS1*, interstage 1; *PA*, pulmonary artery; *BCPC*, bidirectional cavopulmonary connection; *LOS*, length of stay; *TPG*, transpulmonary gradient; *PVR*, pulmonary vascular resistance; *EDP*, end-diastolic pressure; *Qp:Qs*, ratio of pulmonary to systemic blood flow. *pASO vs PAB. †pASO vs Norwood. ‡PAB vs Norwood. §Operative or catheter-based procedures related to cardiac pathology or postsurgical complications.

TABLE E6. Summary of first unplanned interstage intervention

Variable	pASO	Norwood	PAB
Number of patients with unplanned intervention(s) in IS1	10	8	5
First unplanned intervention in IS1	• SPS placement (n = 3) • PAB adjustment (n = 3) • Coronary anastomosis revision (n = 1) • Exploration/mediastinal washout of open chest for bleeding (n = 1) • Incision and drainage for wound infection (n = 1) • Aortic arch balloon angioplasty (n = 1)	• Aortic arch balloon angioplasty (n = 4) • Pacemaker (n = 1) • Incision and drainage for wound infection (n = 1) • LPA stent (n = 1) • SPS upsized + bilateral PA augmentation (n = 1)	• PAB adjustment (n = 1) • Aortic arch balloon angioplasty (n = 1) • Pericardial drain (n = 1) • Balloon atrial septostomy (n = 1) • Pacemaker (n = 1)

pASO, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *IS1*, interstage 1; *SPS*, systemic-to-pulmonary artery shunt; *LPA*, left pulmonary artery.

TABLE E7. Clinical details for postoperative coronary pathology

Pt- initial palliation	Coronary anatomy (baseline)	Postoperative coronary pathology/interventions
1-pASO*	Two coronaries from posterior facing sinuses, RCA with early bifurcation supplying anterior descending and right AV groove/non-dominant circumflex. LCA to L AV groove	Postoperative: intermittent AV block and diffuse ST changes, elevated troponin, wall-motion abnormalities. CTA shows *patent RCA ostium with proximal stenosis*. Concern that dilated neoaortic root placed tension on the sinus **Reoperation (POD 38): RCA dilation and ostial revision**
2-pASO*	LCA from leftward/posterior-facing sinus, continuing as circumflex "RCA" tiny second orifice in anterior cusp with branches to anterior wall of hypoplastic subaortic RV ? Accessory LAD from proximal RCA with attenuated RCA continuation	Intraoperative (index): slow reperfusion, ST changes → coronary anastomosis revision → ST changes improved, continued ventricular dysfunction → ECMO Cath (POD 1): suspicious for *proximal circumflex dissection*, preserved distal flow **Therapeutic anticoagulation only**
3-pASO	Anomalous interarterial + intramural LCA from R aortic sinus of Valsalva (also hypoplastic, ∼1 mm), giving rise to LAD and circumflex RCA juxtacommissural from R sinus	Intraoperative (index): transection and reimplantation of LCA Postoperative: intermittent ST changes Cath (∼2 wk postoperative): *hypoplastic vs stenotic LCA*, deemed unchanged from baseline anatomy noted intraoperatively **No intervention**
4-pASO	Coronary from rightward/anterior sinus → anterior descending + right AV groove to LV; small coronary from leftward/posterior facing sinus in left AV groove	*Iatrogenic injury of acute marginal coronary artery* during Fontan sternotomy at outside hospital—successful **intraoperative repair**
5-Norwood	Inverted pattern from cc-TGA: LCA from right anterior sinus gives LAD and small circumflex, several LAD-RV fistulae. RCA from posterior sinus supplies posterior LV, tortuous course, ∼2 mm (similar size to aorta)	Intraoperative (index): salvage operation for unstable patient as bridge to transplant, septal myectomy and accessory AV valve tissue resection. ECMO for refractory arrhythmias. Postoperative: ECMO decannulated, PM placed for VT and AV block. continued ventricular dysfunction and *concern for coronary steal from fistulae vs strangulation of diagonal branch from ventricular pacing lead* Glenn: **PM wire revision**, also thrombus in shunt Post-Glenn: improved LV function, persistent regional wall motion abnormalities. Completed Fontan, did not require transplant.
6-Norwood	Bilateral coronaries from extreme posterior juxtacommissural position with intramural courses (R > L) and slit-like ostia	Intraoperative (index): no coronary intervention Postoperative: good ventricular function Glenn: ostial enlargement/unroofing of bilateral coronaries Post-Glenn: *severe proximal RCA stenosis* with ventricular dysfunction → **OR translocation of RCA to pulmonary root (neoaorta/DKS)**
7-Norwood	Normal/Yacoub A	Post-Fontan inferolateral ST depression (asymptomatic), stress cardiac MRI with mild inducible perfusion defect in inferior wall and septum. Cath without antegrade perfusion in distal LAD, noted small left coronary system and collateral supply to distal one-third of septum and LAD territory; consistent with *distal CAD post-Fontan*, **no interventions**

All of the patients shown in the table had patient coronaries at subsequent assessment and were alive at follow-up. Italics indicate coronary pathology, bold indicates interventions. *pASO*, Palliative arterial switch operation; *RCA*, right coronary artery; *AV*, atrioventricular; *LCA*, left coronary artery; *CTA*, computed tomography angiography; *POD*, postoperative day; *ECMO*, extracorporeal membrane oxygenation; *LAD*, left anterior descending; *VT*, ventricular tachycardia; *PM*, permanent pacemaker; *LV*, left ventricular; *DKS*, Damus–Kaye–Stansel; *CAD*, coronary artery disease. *Considered potential pASO operative complications.

TABLE E8. PA interventions

Variable	Overall (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB: All (n = 25)	*P* value
Intervention: any time					
Any PA intervention*	48/71 (68%)	18/23 (78%)	15/23 (65%)	15/25 (60%)	.40
Any LPA intervention, n	39	17	13	9	
% total	39/71 (55%)	17/23 (74%)	13/23 (57%)	9/25 (36%)	**.04** ***.01***† *.35*‡ *.25*§
% PA interventions	39/48 (81%)	17/18	13/15	9/15	.06
Any LPA stent, n	8	2	5	1	
% total	8/71 (11%)	2/23 (9%)	5/23 (22%)	1/25 (4%)	.16
% PA interventions	8/48 (17%)	2/18 (11%)	5/15 (33%)	1/15 (7%)	.17
% LPA interventions	8/39 (21%)	2/17 (12%)	5/13 (38%)	1/9 (11%)	.21
Multiple PA interventions	21	11	7	3	
% total	21/71 (30%)	11/23 (48%)	7/23 (30%)	3/25 (12%)	**.03** ***.01***† *.37*‡ *.16*§
% PA interventions	21/48	11/18	7/15	3/15	.06
Imaging: pre-Glenn					
Minimum LPA diameter, mm	4.4 (3.7-5.8) n = 58	4.2 (3-4.9) n = 20	4.4 (4.0-5.5) n = 20	5 (4.1-6.9) n = 18	.11
Minimum LPA diameter/BSA, mm/m^2	14.6 (12.1-17.9)	14.3 (12.0-16.4)	15.0 (12.1-18.2)	14.8 (12.1-18.2)	.65
Minimum RPA diameter, mm	4.5 (3.8-5.8) n = 57	4.8 (4-5.2) n = 20	4.6 (3.8-5.8) n = 19	4.1 (3-7.3) n = 18	.82
Minimum RPA diameter/BSA, mm/m^2	14.9 (11.4-20.8)	16.5 (13.5-21.1)	15.8 (9.7-19.3)	12.1 (8.8-21.1)	.33
McGoon	1.9 (1.6-2.3) n = 18	1.7 (1.6-1.8) n = 5	1.7 (1.5-2.3) n = 6	2.2 (1.9-2.6) n = 7	.11
BCPC					
BCPC PA plasty	39/66 (59%)	16/23 (70%)	12/21 (57%)	11/22 (50%)	.45
Bilateral PAs					
% BCPC PA plasty	24/39 (62%)	12/39 (75%)	7/12 (58%)	5/11 (45%)	.32
% BCPC	24/66 (36%)	12/23 (52%)	7/21 (33%)	5/22 (23%)	.13
LPA‖					
% BCPC PA plasty	28/39 (72%)	14/16 (88%)	8/12 (67%)	6/11 (55%)	.14
% BCPC	28/66 (42%)	14/23 (61%)	8/21 (38%)	6/22 (27%)	.07
RPA‖					
% BCPC PA plasty	34/39 (87%)	14/16 (88%)	11/12 (92%)	9/11 (82%)	.85
% BCPC	34/66 (52%)	14/23 (61%)	11/21 (52%)	9/22 (41%)	.43
Fontan					
Fontan PA plasty	22/55 (40%)	12/19 (63%)	6/18 (33%)	4/18 (22%)	**.04** ***.02***† *.10*‡ *.71*§
Bilateral PA plasty¶					
% Fontan PA plasty	5/22 (23%)	1/12 (8%)	2/6 (33%)	2/4 (50%)	.15
% Fontan	5/55 (9%)	1/19 (5%)	2/18 (11%)	2/18 (11%)	.73

Bold denotes raw *P* value < .05. Post-hoc pairwise comparisons included for significant differences (italics). For Pre-BCPC imaging by cohort and by interventions performed, see Figure E5. *PA*, Pulmonary artery; *pASO*, palliative arterial switch operation; *PAB*, pulmonary artery banding; *PA*, pulmonary artery; *LPA*, left pulmonary artery; *BSA*, body surface area; *RPA*, right pulmonary artery; *BCPC*, bidirectional cavopulmonary connection. *PA interventions defined as operative PA augmentation or catheter-based balloon angioplasty or stent placement. †pASO vs PAB. ‡pASO vs Norwood. §PAB vs Norwood. ‖Includes patients with isolated LPA or RPA plasty as well as part of bilateral branch PA plasty. ¶No patients had isolated RPA plasty at Fontan; all had LPA plasty either isolated (n = 17) or as part of bilateral PA plasty (n = 5).

TABLE E9. Pre-BCPC PA imaging and BCPC intervention

Variable	BCPC LPA plasty n = 28	BCPC no LPA plasty n = 38	*P* value
Min LPA diameter, mm	4.1 (3.5-5) n = 27	4.7 (4-6) n = 31	.18
Min LPA diameter/BSA, mm/m^2	13.5 (11.4-16.9) n = 27	14.6 (12.7-17.9) n = 31	.31
Variable	**BCPC RPA plasty n = 34**	**BCPC no RPA plasty n = 32**	
Min RPA diameter, mm	4.1 (3.5-5) n = 31	5 (4.2-7) n = 26	**.01**
Min RPA diameter/BSA, mm/m^2	14.5 (10.5-16.9) n = 31	17.5 (12.1-22.6) n = 26	**.03**

Bold denotes raw *P* value < .05. See Figure E5, E-H. *BCPC*, Bidirectional cavopulmonary connection; *LPA*, left pulmonary artery; *BSA*, body surface area; *RPA*, right pulmonary artery.

TABLE E10. PA imaging/interventions for anterior-posteriorly oriented great vessels pASO versus Norwood

Variable	pASO + AP (n = 20)	Norwood + AP (n = 19)	*P* value
Pre-BCPC PA plasty	1/20 (5%)	2/19 (11%)	.61
Minimum LPA diameter, mm	4.1 (3-4.8)	4.4 (4-5)	.13
Minimum LPA diameter/BSA, mm/m^2	14.2 (12.4-16.6)	15 (12.8-17.2)	.51
Minimum RPA diameter, mm	4.5 (4-5)	4.3 (3.6-5.1)	.78
Minimum RPA diameter/BSA, mm/m^2	14.8 (13.5-19.3)	15.3 (9.1-17.5)	.33
BCPC	n = 20	n = 18	
BCPC LPA plasty	12/20 (60%)	7/18 (39%)	.33
BCPC RPA plasty	12/20 (60%)	10/18 (56%)	1
Any PA plasty	15/20 (75%)	13/19 (68%)	.73
Any LPA plasty	15/20 (75%)	12/19 (63%)	.50

PA, Pulmonary artery; *AP*, anterior-posterior; *pASO*, palliative arterial switch operation; *BCPC*, bidirectional cavopulmonary connection; *LPA*, left pulmonary artery; *BSA*, body surface area; *RPA*, right pulmonary artery.

TABLE E11. PA imaging/interventions for Norwood patients with anterior-posterior versus side-to-side orientated great vessels

Variable	Norwood + SS (n = 4)	Norwood + AP (n = 19)	*P* value
Pre-BCPC PA plasty	0/4 (0%)	2/19 (11%)	1.0
Minimum LPA diameter, mm	4 (3.7-7)	4.4 (4-5)	.86
Minimum LPA diameter/BSA, mm/m^2	13.2 (11.1-25.3)	15 (12.8-17.2)	.93
Minimum RPA diameter, mm	6 (4.6-8)	4.3 (3.6-5.1)	.13
Minimum RPA diameter/BSA, mm/m^2	21.7 (16.4-22.2)	15.3 (9.1-17.5)	.33
BCPC	n = 3	n = 18	
BCPC LPA plasty	1/3 (33%)	7/18 (39%)	1.0
BCPC RPA plasty	1/3 (33%)	10/18 (56%)	.59
Any PA plasty	2/4 (50%)	13/19 (68%)	.59
Any LPA plasty	1/4 (25%)	12/19 (63%)	.28

SS, Side-side; *AP*, anterior-posterior; *BCPC*, bidirectional cavopulmonary connection; *PA*, pulmonary artery; *LPA*, left pulmonary artery; *BSA*, body surface area; *RPA*, right pulmonary artery.

TABLE E12. Follow-up details and echo data

Variable	Overall (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB (n = 25)	*P* value
Follow-up time (y, all)	8.0 (3.6-16.5) n = 71	7.9 (4.0-16.2)	8.4 (3.6-16.1)	8.0 (3.6-17.5)	.90
Follow-up time (y, survivors)	10.4 (4.5-16.6) n = 66	7.9 (4.0-16.2)	9.8 (4.5-16.6)	13.0 (6.0-17.8)	.50
Lost to follow-up (survivors)	7/66 (11%)	5/23 (22%)	1/21 (5%)	1/22 (5%)	.20
Arrhythmia*					
% total	18/71 (25%)	3/23 (13%)	4/23 (17%)	11/25 (44%)	**.03**
% survivors	16/66 (24%)	3/23 (13%)	4/21 (19%)	9/22 (41%)	.09
Any coarctation reintervention (cath or OR, % total)	9/71 (13%)	2/23 (9%)	5/23 (22%)	2/25 (8%)	.37
Ventricular dysfunction§ (mild or greater)	10/66 (15%)	4/23 (17%)	3/21 (14%)	3/22 (14%)	1.0
Normal-"low-normal"	56/66 (85%)	19/23 (83%)	18/21 (86%)	19/22 (86%)	.81
Mildly depressed	8/66 (12%)	4/23 (17%)	2/21 (10%)	2/22 (9%)	
Moderately depressed	2/66 (3%)	0 (0%)	1/21 (5%)	1/22 (5%)	
Systemic valve insufficiency† (mild or greater)	35/66 (53%)	13/23 (57%)	13/21 (62%)	9/22 (41%)	.37
None-trivial	31/66 (47%)	10/23 (43%)	8/21 (38%)	13/22 (59%)	.15
Mild	32/66 (48%)	10/23 (43%)	13/21 (62%)	9/22 (41%)	
Moderate	3/66 (5%)	3/23 (13%)	0 (0%)	0 (0%)	

Bold denotes raw *P* value <.05. Data provided extracted retrospectively from report of most recent echocardiogram for *survivors*, or last pretransplant echocardiogram for the patient who underwent cardiac transplant. Most severe qualitative measure was used, when ranges were reported. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *OR*, operating room; *DKS*, Damus–Kaye–Stansel. *Arrhythmia requiring antiarrhythmic medication or pacemaker at latest follow-up. †Systemic valve insufficiency included neoaortic valve insufficiency for those with pASO or DKS and native aortic valve insufficiency for patients with PAB without DKS. Presence of systemic valve insufficiency defined as qualitatively mild or greater. §Presence of ventricular dysfunction defined as qualitatively mild or greater ventricular dysfunction (low-normal function was not considered ventricular dysfunction).

TABLE E13. Systemic valve insufficiency by PAB subgroups (with vs without DKS)

Variable	Overall (n = 71)	pASO (n = 23)	Norwood (n = 23)	PAB:+ DKS (n = 13)	PAB: no DKS (n = 12)	*P* value
Systemic valve insufficiency* (mild or greater)	35/66 (53%)	13/23 (57%)	13/21 (62%)	5/12 (42%)	4/10 (40%)	.56
None-trivial	31/66 (47%)	10/23 (43%)	8/21 (38%)	7/12 (58%)	6/10 (60%)	.43
Mild	32/66 (48%)	10/23 (43%)	13/21 (62%)	5/12 (42%)	4/10 (40%)	
Moderate	3/66 (5%)	3/23 (13%)	0 (0%)	0 (0%)	0 (0%)	

Data provided extracted retrospectively from report of most recent echocardiogram for survivors, or last pre-transplant echocardiogram for the patient who underwent cardiac transplant. Most severe qualitative measure was used, when ranges were reported. *pASO*, Palliative arterial switch operation; *PAB*, pulmonary artery banding; *DKS*, Damus–Kaye–Stansel. *Systemic valve insufficiency included neo-aortic valve insufficiency for those with pASO or DKS and native aortic valve insufficiency for PAB patients without DKS. Presence of systemic valve insufficiency defined as qualitatively mild or greater.