Simultaneous Systemic to Pulmonary Shunt and Pulmonary Artery Banding is a Viable Option for Neonatal Palliation of Single Ventricle Physiology

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A subset of neonates with single ventricle (SV) physiology has antegrade pulmonary blood flow that is deemed unlikely to be reliable until Glenn. We have used systemic to pulmonary shunt (SPS) with pulmonary artery banding (PAB) to optimize pulmonary blood flow while maintaining reserve antegrade flow. We hypothesize that this is an effective strategy that can be accomplished without the routine need for cardiopulmonary bypass. We retrospectively reviewed the records of 60 neonates who underwent combined SPS + PAB between 2004 and 2015. Data are presented as median with quartiles. Children were 8 (4–19) days old at surgery and included 38 (63%) boys. Atresia or severe stenosis of the subpulmonary atrioventricular (AV) valve associated with pulmonary blood flow across a bulboventricular foramen was present in 37 (62%). In 20 (33%), heterotaxy-associated unbalanced AV canal with pulmonary stenosis with or without anomalous pulmonary venous drainage was present. First-stage palliation was accomplished without cardiopulmonary bypass in 44 patients (73%). There were 7 (12%) hospital deaths, 4 among the 20 (20%) with heterotaxy. Fifty-three children were followed for a median 5.1 (1.8–8.2) years. Three early reinterventions were required after initial palliation (1 PAB adjustment, 2 SPS balloon angioplasties). Five additional heterotaxy patients experienced late mortality during follow-up. There were no early or emergent Glenn. Thirty-nine patients have reached Fontan circulation with a median pre-Fontan PA pressure of 14 (12–18) mm Hg. One patient converted to biventricular physiology and the remaining await completion Fontan. Heterotaxy was the only independent predictor of mortality (hazard ratio 10 (2.3–44, $P < 0.001$). In SV patients with unreliable antegrade PA flow, SPS + PAB is an effective first-stage palliation. SV patients with heterotaxy are at increased risk for mortality.

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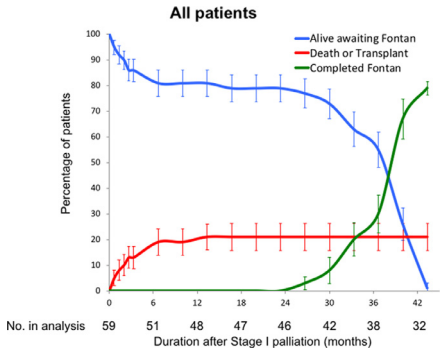
Keywords: Single ventricle, Heterotaxy, Shunt, PA band, Fontan



Competing outcomes analysis in SV patients undergoing neonatal SPS + PAB.

Central Message

In single ventricle patients with unreliable pulmonary blood flow, simultaneous systemic to pulmonary shunt and pulmonary artery banding is a viable option for neonatal palliation.

Perspective Statement

A subset of single ventricle patients presents with variable obstruction to pulmonary blood flow that is deemed unreliable until progression to Glenn. Simultaneous systemic to pulmonary shunting and pulmonary artery banding is an acceptable first-stage palliation in these patients and results in excellent single ventricle outcomes. Heterotaxy-related defects represent an increased risk for mortality.

Abbreviations: AV, atrioventricular; BT, Blalock-Taussig; BVF, bulboventricular foramen; CPB, cardio-pulmonary bypass; DILV, double inlet left ventricle; DORV, double outlet right ventricle; ECMO, extracorporeal membrane oxygenator; EF, ejection fraction; NYHA, New York Heart Association; PAB, pulmonary artery band; PBF, pulmonary blood flow; SPS, systemic-pulmonary shunt; SV, single ventricle; d-TGA, dextro-transposition of great arteries; VSD, ventricular septal defect

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BACKGROUND

The surgical management of neonates born with single ventricle (SV) physiology includes a series of staged procedures leading to Fontan circulation. Successful establishment of Fontan circulation begins with the appropriate neonatal surgical approach. The major goals of the neonatal procedure are to establish unobstructed systemic outflow, adequate pulmonary blood flow (PBF), and unrestricted mixing of blood in the heart.[1]

It is well recognized that patients with SV physiology present with variable degrees of native PBF. At one end of the spectrum are patients with pulmonary atresia whose PBF is entirely ductal-dependent. In this subset, neonatal palliation usually requires placement of a systemic to pulmonary shunt (SPS).[2] At the other end of the spectrum are children with abundant, unrestricted PBF who require pulmonary artery banding (PAB) to appropriately restrict PBF.[3] One subset of patients presents with varying degrees of obstruction to PBF caused by restriction through a bulboventricular foramen (BVF) or due to valvar or subvalvar pulmonary stenosis causing unpredictable PBF. Appropriate neonatal palliation that ensures adequate saturations while preventing pulmonary overcirculation can be challenging in this group of patients. We have used SPS with simultaneous PAB in these patients to ensure adequate PBF while maintaining reserve antegrade flow. This study aims to report our institutional experience with this management strategy.

METHODS

At our institution, 742 patients underwent first-stage SV palliation between 2004 and 2015. Under institutional review board-approved protocols, we retrospectively reviewed the charts of 60 consecutive patients in this cohort who underwent simultaneous SPS and PAB to address unreliable PBF. Data on demographics, SV morphology, and diagnostic investigations in the neonatal period were collected. During this same time period, 381 patients with entirely ductal-dependent PBF underwent isolated SPS and 44 patients with unrestricted excess PBF underwent isolated PAB placement. Outcomes in these patients are not included in the current analysis.

Our institutional preference is to approach these lesions via median sternotomy. Cardiopulmonary bypass (CPB) is used selectively in those patients who require additional procedures that need bypass or experience hemodynamic instability when pulmonary artery is clamped for distal shunt anastomosis. We initially place an SPS. In the majority of instances, this was done using a modified Blalock-Taussig (BT) shunt (3.5—4.0 mm). The size of the shunt is chosen largely based on patient weight—we tend to use 3.5 mm shunt in patients weighing up to about 3 kg. The ductus is then ligated and any other anomaly requiring correction is addressed. A thin silastic PA band is then placed around the main PA and secured to the right ventricular outflow tract to prevent impingement on branch PAs. The band is tightened to allow antegrade PBF that leads to systemic saturations in the 85—90% range. We have found that this results in a main PA size that is slightly smaller than the size of the aorta. Intraoperative transesophageal echocardiography is utilized to quantify gradients across the band by Doppler interrogation. We generally aim for a peak Doppler velocity between 3 and 4 m/s across the band. Operative reports were reviewed to obtain CPB times and need for cardiac and circulatory arrest.

Subsequent stages of SV palliation followed current standard practice patterns. Patients are followed by serial echocardiography and undergo routine presurgical cardiac catheterization. Figure 1 and Video 1 show the amount of antegrade PBF in a representative patient just prior to Glenn and Fontan. At 6—8 months of age, bidirectional superior cavopulmonary anastomosis (Glenn procedure) is performed under CPB and the shunt is ligated. In the majority of patients, the band is further tightened such that the pressure waveform in the Glenn circuit is nonpulsatile. This results in a PA approximately 2—3 mm in diameter that carries a small amount of residual antegrade blood flow. In the rest of the patients, based on individual anatomy, the main pulmonary artery is divided and the valve excised. At about 3 years of age, total cavopulmonary connection (Fontan procedure) is established with an extracardiac Goretex conduit and selectively fenestrated. For this study, interstage data leading to the Glenn procedure and ultimate SV outcomes were reviewed.

Data were tabulated and analyzed using SAS 9.4. Continuous data are presented as median with quartiles; discrete data are presented as number with percentages. Competing outcomes analysis was undertaken to study overall progression to Fontan and cumulative incidence function analysis used to quantify the impact of heterotaxy. Cox regression analysis was

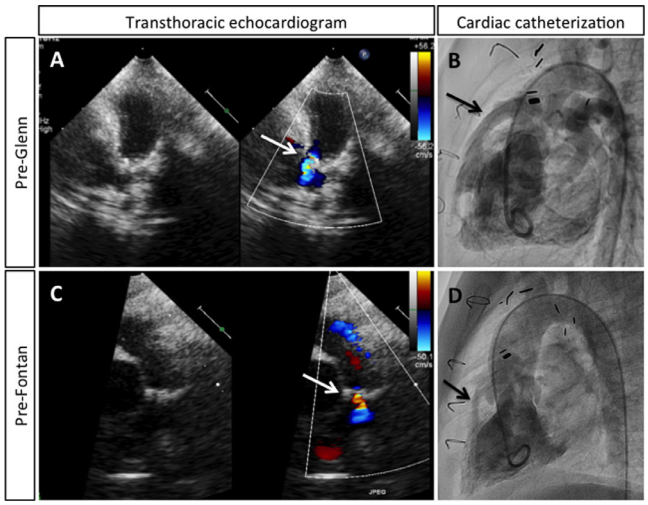


Figure 1. Representative transthoracic short-axis view (right panel is color Doppler image of the left panel) of right ventricular outflow before Glenn (A) and Fontan (C) in the same patient. Antegrade PBF across PAB is indicated by white arrow. Corresponding ventriculograms (B and D) during cardiac catheterization also demonstrating the amount of antegrade PBF (black arrow).

used to assess predictors of mortality. Significance was defined as $P < 0.05$.

RESULTS

Patient Demographics

Characteristics of the 60 patients who underwent simultaneous SPS and PAB are shown in Table 1. There were 2 predominant SV morphologies. Thirty-seven (62%) patients had atresia of the subpulmonary atrioventricular (AV) valve associated with PBF across a BVF. Of these, 19 patients had tricuspid atresia with a restrictive BVF, normally related great vessels and variable degree of pulmonary stenosis. Three patients had double inlet left ventricles and a similarly restrictive BVF with/without right ventricle outflow tract obstruction. An additional 15 patients had left AV valve atresia in the setting of malposed or transposed vessels such that antegrade PBF relied on a restrictive ventricular septal defect (VSD). The second most common morphology was heterotaxy-associated unbalanced AV canal defect, and double outlet right ventricle with pulmonary stenosis in 20 (33%) patients. Nine of these patients also had anomalous pulmonary venous drainage. The remaining 3 patients had anomalies in great vessel orientation (DORV or d-TGA), with remote VSD and small right-sided structures.

Neonatal Palliation

Operative details of neonatal palliation are shown in Table 2. Fifty-seven percent patients received a 4 mm BT shunt. Data on peak PAB gradient on intraoperative transesophageal echocardiography were available in 49 patients. The median peak gradient in these patients was 48 mm Hg (36–59.75). In 73% patients, SPS and PAB were completed without the use of CPB. Of the remaining 16 patients, 6 required circulatory arrest—4 for repair of pulmonary venous return and 2 for aortic arch augmentation. Four patients required aortic crossclamping for atrial septectomy (3)[3] or closure of a regurgitant and stenotic AV valve (1).[1] Three patients required unifocalization and/or augmentation of the PA under CPB. Four patients required CPB for hemodynamic instability during off-pump SPS construction. Of the 16 patients who required CPB, 8 (50%) had heterotaxy.

Morbidity was assessed based on Society of Thoracic Surgeons database criteria (Table 2). Four patients required extracorporeal membrane oxygenation (ECMO) support in the perioperative period. This included 1 patient with heterotaxy who developed significant dysrhythmia and hemodynamic compromise 16 hours after surgery. Another patient with heterotaxy and severe AV valve regurgitation had inadequate cardiac output and could not be weaned from CPB. A third heterotaxy patient with obstructed TAPVR who underwent emergent palliation had persistent inadequate arterial saturations despite an adequate shunt and TAPVR repair. His pulmonary artery was debanded to allow more antegrade blood flow, and yet, did not have adequate PBF. The fourth patient developed sudden profound desaturation followed by circulatory collapse 8 hours after surgery. He was emergently placed on ECMO but developed massive intracranial bleed within the next few hours precluding further evaluation. Two patients required reoperation for loosening of PA band due to inadequate PBF despite having a 4 mm BT shunt that was felt to be adequate and patent. Both these patients ultimately expired

Table 1. Patient Characteristics ($n = 60$)

Male sex	37 (62)
Race	
Hispanic	30 (50)
Caucasian	17 (28)
African-American	9 (15)
Other	4 (7)
Prematurity	3 (5)
Genetic syndrome	2 (3.3)
Heterotaxy	20 (33)
Single ventricle morphology	
Tricuspid valve atresia or DILV and VSD	22 (37)
Unbalanced AV canal/DORV	20 (33)
Mitral atresia with malposed or transposed vessels	15 (25)
Other	3 (5)
Prostaglandin-dependent pulmonary circulation	11 (18)

Data are represented as number (percentage).
AV, atrioventricular; DILV, double inlet left ventricle; DORV, double outlet right ventricle; VSD, ventricular septal defect.

Table 2. Operative Characteristics at First Stage Palliation ($n = 60$)

Weight (kg)*	3.21 (2.8–3.7)
Age (d)*	8 (4–19)
Systemic to pulmonary shunt	
Modified Blalock-Taussig (3.5 mm)	24 (40)
Modified Blalock-Taussig (4 mm)	34 (57)
Classic shunt	2 (3)
Pulmonary artery band peak gradient (mm Hg)*,†	48 (36–60)
No. requiring cardiopulmonary bypass	16 (27)
Duration of bypass (min)*	37 (31–42)
No. requiring cardiac arrest	9 (15)
Duration of arrest (min)*	24 (23–27)
No. requiring circulatory arrest	6 (10)
Duration of arrest (min)*	21 (18–24)
Cardiac-related morbidity events‡	9 (15)
Need for ECMO	4 (7)
Unplanned band adjustment	2 (3)
Shunt revision	2 (3)
Other	5 (8)
Mortality	7 (12)
Duration of postoperative hospital stay (d)*	21 (18–31)

ECMO, extracorporeal membrane oxygenation.
*Data are presented as median and quartiles. Rest of data are number (percentage).
†Based on data from 49 patients.
‡Not mutually exclusive.

Table 3. Causes of Mortality

Mortality during first-stage palliation
Inadequate pulmonary blood flow in a patient with obstructed TAPVR at birth (heterotaxy)
Malignant dysrhythmia leading to hemodynamic compromise (heterotaxy)
Severe AV valve regurgitation and persistent dysrhythmia (heterotaxy)
Abdominal complications and subsequent septic shock (heterotaxy)
Unexplained cardiac arrest and subsequent intracranial bleed
Chronic respiratory failure and sepsis
Aspiration leading to pneumonia and overwhelming sepsis
Mortality between first and second stage (all heterotaxy)
Severe AV valve regurgitation and severe ventricular dysfunction (2)[2]
Abdominal complications requiring bowel resection, subsequent sepsis (2)[2]
Recurrent obstruction at TAPVR repair site, with diffuse pulmonary vein stenosis

during the hospitalization. Two BT shunts developed thrombosis in the immediate postoperative period and were revised to a classic shunt in 1 and a central shunt in the other. There were 5 other minor morbidity events—3 superficial wound infections, 1 exploration for bleeding, and 1 pericardial effusion drainage. There were 7 (12%) hospital mortalities, 4 (20%) of who had heterotaxy syndrome (Table 3).

Table 4. Progress to Second-Stage Palliation Among 53 Survivors

No. of patients with at least 1 readmission	14
No. of patients with more than 1 readmission	4
Reason for readmission*	
Inadequate arterial saturation	8
Gastrointestinal issues	8
Dysrhythmia	2
Other	3
Unplanned cardiac reinterventions	
Band adjustment	1 (2)
Shunt angioplasty	2 (4)
Mortality	5 (9)
Age at Glenn (mo, 48 patients)†	7.2 (5.4–8.2)
Concomitant procedures at Glenn (48 patients)*	20 (42)
Interruption of antegrade pulmonary blood flow	12 (25)
Atrial septectomy	5 (10)
Pulmonary arterioplasty	4 (8)
Other	2 (4)

*Not mutually exclusive.
†Data are presented as median and quartiles. Rest of data are number (percentage).

Interstage Events

The subsequent course in the 53 patients who survived to discharge following neonatal palliation was then studied. Events leading up to their Glenn procedure are shown in Table 4. Fourteen patients required 21 readmissions. Eight of these were for inadequate arterial saturations. In 6 patients, this was thought to be related to intercurrent illnesses and did not require reintervention. Two patients had SPS stenoses requiring balloon angioplasty. Two patients with heterotaxy were admitted for dysrhythmia management. Eight patients had gastrointestinal issues such as feeding intolerance (one secondary to adhesive bowel obstruction related to prior neonatal Ladd's), necrotizing enterocolitis, and diarrhea requiring admission. This includes 3 heterotaxy patients who required Ladd's procedure (2)[2] or bowel resection (1).[1] One patient required PAB tightening for persistent pulmonary overcirculation and high saturations. Five patients, all with heterotaxy, died during the interstage period (Table 3). Two of these patients developed severe AV valve regurgitation not amenable to repair and severe ventricular dysfunction, 1 had obstruction at site of repair of anomalous pulmonary venous return with diffuse pulmonary venous stenosis, and 2 developed intestinal complications.

The 48 survivors of the interstage period underwent Glenn procedure at median of 7.2 (5.4–8.2) months of age. Twenty patients required 23 concomitant procedures during Glenn (Table 4). Five patients required atrial septectomy, and each one required revision of pulmonary venous repair and pacemaker implantation. Our usual approach at this stage would be to further tighten the PAB to maintain a small amount of antegrade PBF. In 12 patients, the antegrade flow was interrupted by pulmonary valvectomy and main PA division. Four of these patients needed extensive PA plasty and 5 other patients required bilateral Glenn. In each case, division of the MPA was helpful in PA reconstruction and/or placement of Glenn anastomosis.

Single Ventricle Outcomes in Patient Cohort

One patient with a complex transposition anatomy was converted to biventricular circulation following neonatal SPS + PAB. We performed competing outcomes analysis in the remaining 59 patients, stratified by the presence or absence of heterotaxy syndrome (Fig. 2). Twelve (20%) patients did not survive to Fontan circulation (Table 5). Figure 2 demonstrates that the mortality curve has a steep early phase and becomes flat at roughly 8 months of age. This is concordant with the observation that all deaths occurred either during neonatal palliation or in the interstage period. Of these 12 deaths, 9 (75%) had heterotaxy syndrome. Cox regression analysis showed that heterotaxy was an independent predictor of mortality with an odds ratio of 10 (2.3–44, $P = 0.0006$).

Of the survivors, 2 patients are awaiting Glenn and 6 are awaiting Fontan (Table 5). Thirty-nine patients have reached Fontan circulation. Mean PA pressure prior to Fontan was 14 mm Hg in these 39 patients, indicating favorable

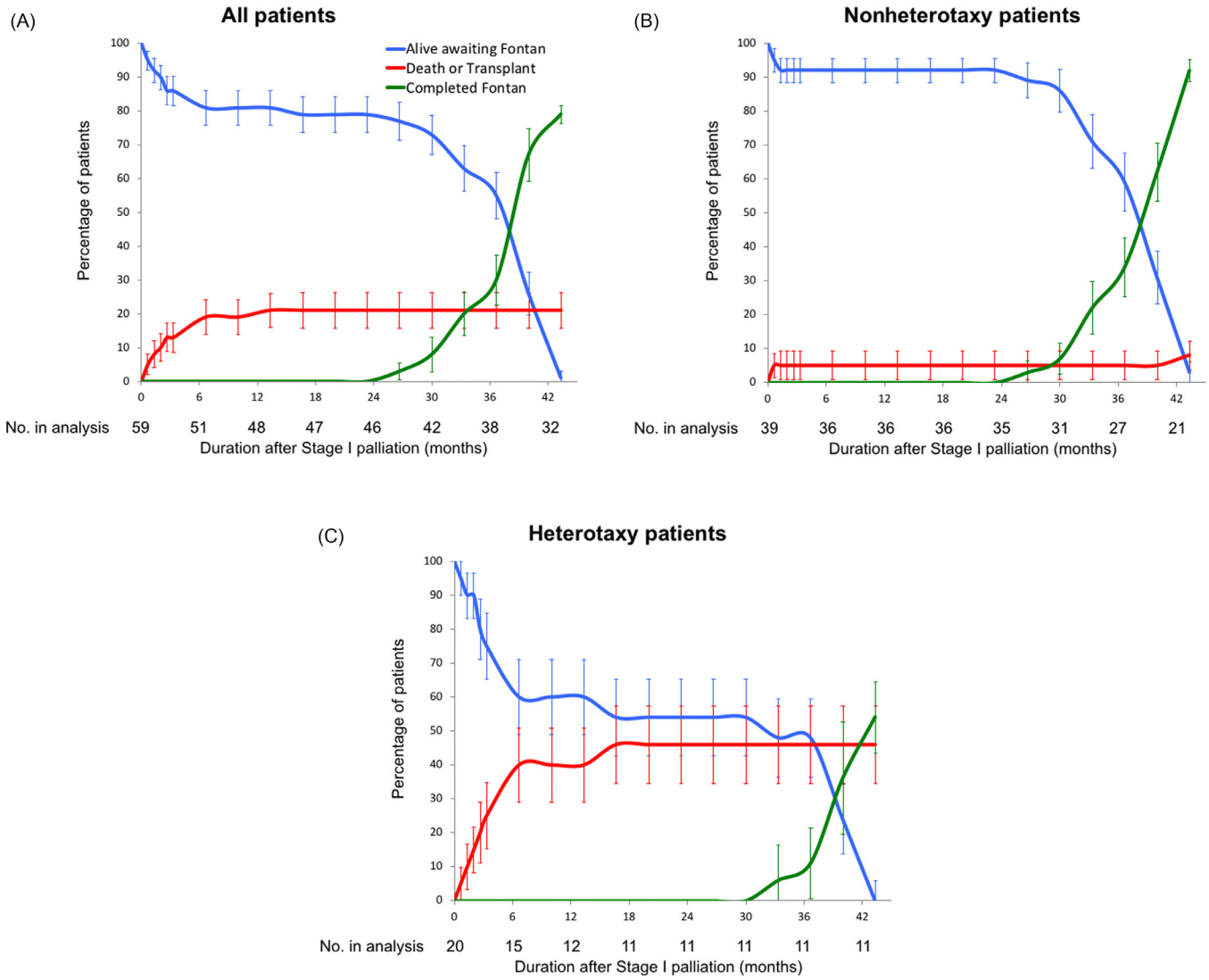


Figure 2. Competing outcomes analysis in the 59 patients who underwent SV palliation (A), stratified as patients without (B) and with (C) heterotaxy syndrome. Error bars indicate 95% confidence limits.

Table 5. Patient Outcomes ($n = 60$)

Died	12 (20)
Hospital mortality during first-stage palliation	7 (12)
Median age at death (d)*	26 (11–49)
Mortality between first- and second-stage palliation	5 (8)
Median age at death (d)*	142 (138–157)
Converted to biventricular circulation	1 (2)
Awaiting Glenn	2 (3)
Awaiting Fontan	6 (10)
Reached Fontan circulation	39 (65)
Pre-Fontan mean pulmonary artery pressure (mm Hg)*	14 (12–18)
Median age at Fontan (mo)*	38 (33–45)

*Data are presented as median and quartiles. Rest of data are number (percentage).

pulmonary hemodynamics for successful Fontan completion. No patient underwent fenestration of Fontan circuit. At a median follow-up of 5.1 (1.8–8.2) years, there was no additional mortality or a need for transplantation. As in the case of mortality, cumulative incidence function analysis (Fig. 3) demonstrated that heterotaxy syndrome was associated with 3.2-fold (1.5–6.9, $P = 0.0016$) reduction in likelihood of reaching Fontan circulation. We collected data from the last clinic visit and surveillance echocardiography to document the clinical status of the patients. Thirty-eight patients were asymptomatic; 1 develops shortness of breath with exertion. Median oxygen saturation was 97% (94–98). None of the patients was on supplemental oxygen or pulmonary vasodilator therapy. Thirty-four patients had normal ventricular function, 5 had mild dysfunction. Median EF at rest was 64% (52–74).

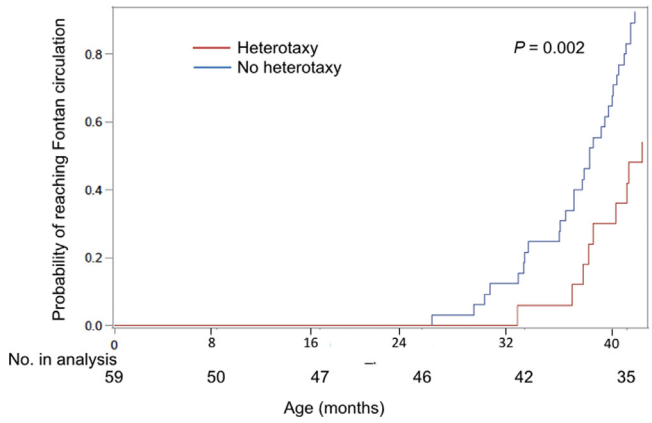


Figure 3. Cumulative incidence function analysis of likelihood of reaching Fontan circulation in patients with and without heterotaxy syndrome. Error bars are shown in Figure 2B and C Fontan curves.

DISCUSSION

Favorable pulmonary hemodynamics plays an important role in achieving successful Fontan circulation. Appropriate management of PBF, starting with neonatal palliation, can positively influence pulmonary vascular maturation. While PBF management may be straightforward in many SV patients, we believe that a subset of neonates has unreliable PBF. These patients have antegrade PBF, which, while not entirely insignificant, is deemed unlikely to be reliable up to the time of second-stage palliation. Our current study relates to this cohort of patients and comprises 2 main anatomic subtypes of SV physiology. Children with atresia of the subpulmonary AV valve (tricuspid atresia, normally related great vessels or mitral atresia, malposed great arteries) and a VSD represent the first subtype. Pulmonary circulation is reliant on flow across the BVF that may not be of adequate size, or be muscle-bound and at risk of progressive obstruction over time. In addition, these children frequently have associated subvalvar or valvar pulmonary stenosis, which can cause dynamic obstruction that may worsen with time. The second anatomic subtype in our study subjects is heterotaxy-related unbalanced AV canal with small right-sided structures, and variable obstruction to right ventricular outflow.

There are several potential approaches to manage PBF in these patients. The addition of an appropriate-sized SPS (or its equivalent, such as a ductal stent) would ensure adequate saturations until second-stage palliation. Several reports in the literature have evaluated the outcomes of a modified SPS in neonates for SV palliation. In 174 patients treated over an 11-year period, Alsoufi et al[4] report 15% hospital mortality and 27% attrition prior to Glenn procedure. There were 14% unplanned reoperations, including 10 shunt revisions. McKenzie et al studied 471 SV patients who underwent SPS over a 17-year period.[5] There were 4% unplanned reinterventions and 15% hospital mortality. Bove et al[6] report a similar 13% mortality in SV patients who receive an SPS, and an 18% shunt-related complication. We believe that addition of an SPS with retention of native antegrade PBF runs the risk of pulmonary overcirculation, with its attendant undesirable hemodynamic consequences. In support of this hypothesis, prior reports of isolated SPS[4] have included patients who required subsequent PAB to limit PBF. In addition, it has been suggested[7] that the presence of unrestricted antegrade PBF is associated with a trend toward increased SPS thrombosis and mortality. A second option to manage our cohort of patients would be to band the main PA alone with the plan to convert to an early Glenn should severe cyanosis ensue. Alsoufi et al reviewed their outcomes of PAB in 73 SV patients over an 11-year period.[3] They report 22% unplanned reoperations, including 5 patients who required band adjustment and 7 patients who required addition of an SPS for inadequate PBF. An earlier study by Rodefeld et al[8] revealed a 5% mortality and 2.5% need for band adjustment in 80 SV patients who underwent PAB. Twenty-one percent of these patients required subsequent pulmonary arterioplasty. We believe that the unpredictable nature of the subpulmonary elements makes banding alone potentially unreliable in our cohort of patients. Pulmonary overcirculation is more easily managed than cyanosis. If severe cyanosis were to ensue, some children may need to be prematurely transitioned to Glenn circulation. We believe that draining the cerebral venous return into an underdeveloped pulmonary vascular tree could result in cerebral venous hypertension and adversely impact neurologic development.

For these reasons, our bias under these circumstances has been to perform a combination of SPS and PAB. We construct an SPS of appropriate size for the child's body surface area to provide adequate PBF, with a tight PAB to provide restricted reserve antegrade flow. We believe that this approach provides more reliable PBF and offers specific advantages. An adequate size SPS, which will ensure dependable PBF until second-stage palliation, can be reliably established in most cases. The addition of a small amount of antegrade flow provides a cushion should there be a compromise to shunt flow. Furthermore, by restricting the amount of antegrade PBF, we prevent pulmonary overcirculation and mitigate the potential problem of competitive flow. There are conflicting reports on the outcomes of complete interruption of antegrade PBF at the neonatal stage.[9,10] In 6 of 22 patients in their series, Bradley et al interrupted antegrade PBF and established an SPS as the sole source of PBF.[9] They suggest that this may be the preferred strategy for neonatal palliation in children with unrestricted PBF. In contrast, a recent report by Wilder et al implies that concomitant main PA intervention at the time of SPS for neonatal SV palliation results in increased mortality.[10] However, of the 25 patients who were included in this analysis, only 4 had PAB, with the remainder having MPA division. To most effectively eliminate antegrade PBF and the risk of systemic embolism, main PA division and pulmonary valvectomy under CPB would eventually be required. However, we believe that this might be best deferred. Only 27% of our entire cohort required CPB at first-stage palliation and among patients who

underwent SPS and PAB alone, only 4 required CPB. While neonatal CPB continues to get safer, there are intuitive benefits to avoiding it when feasible.[11,12] A second potential advantage to maintaining antegrade PBF through to the Fontan stage relates to the continuing supply of hepatic factors to the pulmonary circulation. It has been suggested[13–15] that lack of hepatic growth factors reaching the pulmonary vasculature is an important etiologic factor for pulmonary arteriovenous malformations. We therefore tighten the PAB at the time of Glenn leaving a small amount of flow and completely interrupt the MPA and remove the pulmonary valve leaflets during Fontan procedure. That said 12 patients in our group had the main pulmonary artery divided at the time of Glenn, precluding them of the potential benefit of hepatic factor supply to the lungs. The 2 most common reasons to divide the main PA were the need for wide PA mobilization for PA plasty and/or optimizing PA orientation for bilateral bidirectional Glenns.

Because of a long-standing institutional bias toward managing SV patients with unreliable PBF by combined SPS and PAB, we do not have a comparable cohort of patients with similar physiology managed by an alternative strategy at our institution. Therefore, the primary aim of our current work is to report the feasibility of combined SPS and PAB as a viable option in this patient population. Our results show that SPS and PAB can be achieved with excellent short-term results. Our 12% mortality compares favorably to published reports of SPS palliation in SV neonates.[4,5] Our band adjustment rate is also comparable to prior reports of PAB in SV patients.[3,8] We speculate that the low 3% SPS reintervention rate seen in our population may be related to the protective effect of supplemental antegrade PBF. Only 4 patients subsequently required augmentation of the pulmonary arteries, 2 of who underwent neonatal pulmonary artery intervention in the form of unifocalization in one, and augmentation of hypoplastic vessel in the other. This indicates that the combination of SPS and PAB does not pose an unusual risk to continued pulmonary artery growth. Competing outcomes analysis shows that the mortality curve is flat following conversion to stage II physiology. Surviving patients are excellent Fontan candidates with mean PA pressures of 14 mm Hg, implying that our approach favorably remodels the pulmonary vasculature for successful Fontan conversion.

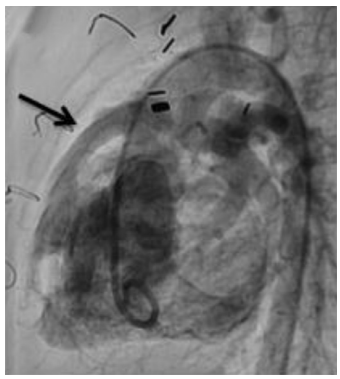
As reported in prior studies,[16–19] heterotaxy was an independent predictor of poor outcome. The anatomic substrate was high risk, with many patients having bilateral cavae and associated pulmonary venous abnormalities. Heterotaxy was not only an independent predictor of mortality, but also predicted inability to reach Fontan circulation. In the neonatal period, increased mortality (20%) may be a reflection of complex anatomic derangements. In particular, associated pulmonary venous anomalies put them at high risk for increased pulmonary vascular resistance and reduced PBF. In the case of the 2 patients who had adequate size SPS and yet required PAB loosening to improve saturations, we suspect that they had high pulmonary resistance that precluded adequate PBF, eventually resulting in death. Following successful neonatal palliation, heterotaxy patients continue to have a higher risk of mortality before Glenn (31%). This is related to progressive AV valve regurgitation and ventricular dysfunction, and to noncardiac abdominal complications. Interestingly, heterotaxy patients who reach Glenn progress through Fontan without any additional mortality risk.

Our study suffers from limitations inherent to a single-institution retrospective study. Strong institutional bias in managing this physiology is inevitable. Because of our universal policy of managing all SV patients whose PBF is thought to be unreliable by combined SPS and PAB, a true institutional control group to compare our results does not exist. We have therefore resorted to comparing our results to published literature. In the absence of a multi-institutional randomized study comparing the various strategies to manage this cohort of patients, we do not believe that a conclusion on the most favorable approach can be reached. Second, this study relates to a relatively small group of patients chosen based on their surgical approach rather than as a disease-based inception cohort. Finally, the long duration of the study may have introduced era-effects that cannot be evaluated in the current study.

Our results show that a combination of systemic to pulmonary shunting and PAB is an acceptable first-stage palliation in patients with SV physiology and unreliable PBF. This can be accomplished without the need for CPB in the majority of cases. Such a strategy results in SV outcomes and long-term physiology that are comparable to published results in such a patient population. Heterotaxy-related SV defects represent an increased risk for mortality and failed conversion to Fontan circulation.

SUPPLEMENTARY MATERIAL

The following is the supplementary data to this article:



Videos 1A–1D. Representative transthoracic short-axis clip (right panel is color Doppler image of the left panel) of right ventricular outflow before Glenn (A) and Fontan (C) in the same patient. Corresponding ventriculograms (B and D) during cardiac catheterization also demonstrating the amount of antegrade PBF.

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