Surgical management of competing pulmonary blood flow affects survival before Fontan/Kreutzer completion in patients with tricuspid atresia type I

Travis J. Wilder, MD,[a] Gerhard Ziemer, MD, PhD,[b] Edward J. Hickey, MD,[c] Peter J. Gruber, MD, PhD,[d] Tara Karamlou, MD, MSc,[e] Paul M. Kirshbom, MD,[f] Eugene H. Blackstone, MD,[g] William M. DeCampli, MD, PhD,[h] William G. Williams, MD,[a] and Brian W. McCrindle, MD, MPH[i]

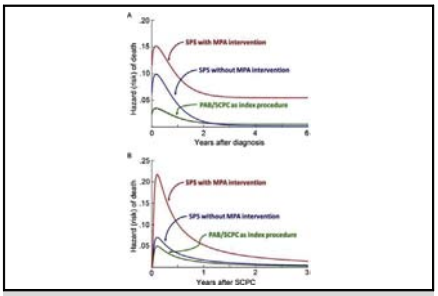
ABSTRACT

Objectives: To determine the association between surgical management of pulmonary blood flow (PBF) at initial and staged procedures with survival to Fontan/Kreutzer operation (Fontan) in patients with tricuspid atresia.

Methods: Infants aged <3 months with tricuspid atresia type I (n = 303) were enrolled from 34 institutions (1999-2013). Among those who underwent surgical intervention (n = 302), initial procedures were: systemic to pulmonary artery shunt (SPS; n = 189; 62%); pulmonary artery banding (PAB; n = 50; 17%); and superior cavopulmonary connection (SCPC; n = 63; 21%). Multiphase parametric-hazard models were used to analyze competing outcomes.

Results: Risk-adjusted 6-year survival was lower after SPS (85%; $P = .04$) versus PAB (93%) or SCPC (93%). Survival after SPS when the main pulmonary artery (MPA) was closed (n = 21) or banded (n = 4) was 60%, versus 93% without MPA intervention ($P = .02$). After SPS, survival before SCPC was lower with an open ductus arteriosus (n = 7; 76% vs 97%; $P = .02$). Similarly, after SPS, risk-adjusted survival was similar to that for patients who had an initial PAB or SCPC when MPA intervention was avoided and the ductus arteriosus either closed spontaneously before SPS, or was closed during SPS. For all patients reaching SCPC (n = 277), survival to Fontan was not significantly influenced by whether PBF persisted through the MPA.

Conclusions: Tricuspid atresia patients with SPS represent a high-risk subgroup. Avoiding an open ductus arteriosus and concomitant MPA intervention during SPS may help mitigate the risk associated with SPS. The presence of antegrade PBF through the MPA, at initial and staged operations, did not significantly influence survival to Fontan operation. (J Thorac Cardiovasc Surg 2015;■:1-9)



The early and late impact on death of main pulmonary artery interruption during an initial SPS procedure. *SPS*, Systemic to pulmonary artery shunt; *MPA*, main pulmonary artery; *PAB*, pulmonary artery band; *SCPC*, superior cavopulmonary connection.

Central Message

For children with tricuspid atresia, judicious management of competing pulmonary blood flow may improve survival to Fontan operation.

Perspective

Infants with tricuspid atresia undergoing initial SPS procedures represent a high-risk subgroup. Avoiding an open ductus arteriosus and concomitant MPA intervention during SPS may help mitigate the risk associated with SPS. The presence of antegrade PBF through the MPA, at initial and staged operations, did not significantly influence survival to Fontan palliation.

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From the [a]Congenital Heart Surgeons' Society Data Center, The Hospital for Sick Children, Toronto, Ontario, Canada; [b]Department of Surgery, Division of Cardiac & Thoracic Surgery, University of Chicago Medicine, Chicago, Ill; [c]Department of Cardiovascular Surgery, University of Toronto, The Hospital for Sick Children, Toronto, Ontario, Canada; [d]University of Iowa Carver College of Medicine, Iowa City, Iowa; [e]Division of Pediatric Cardiac Surgery, Benioff Children's Hospital, University of California San Francisco, San Francisco, Calif; [f]Division of Cardiothoracic Surgery, Connecticut Children's Medical Center and Yale-New Haven Children's Hospital, Hartford and New Haven, Conn; [g]Heart and Vascular Institute, Cleveland Clinic, Cleveland, Ohio; [h]Department of Cardiothoracic Surgery, Arnold Palmer Hospital for Children, Orlando, Fla; and [i]Division of Pediatric Cardiology, Department of Pediatrics, University of Toronto and The Hospital for Sick Children, Toronto, Canada.

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Address for reprints: Brian W. McCrindle, MD, MPH, The Hospital for Sick Children, 555 University Ave, Toronto, Ontario M5G 1X8, Canada (E-mail: brian.mccrindle@sickkids.ca).

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Abbreviations and Acronyms
CHSS = Congenital Heart Surgeons' Society
MPA = main pulmonary artery
PAB = pulmonary artery banding
PBF = pulmonary blood flow
PDA = patent ductus arteriosus
SCPC = superior cavopulmonary connection
SPS = systemic to pulmonary artery shunt

Supplemental material is available online.

Survival of patients with tricuspid atresia and normally related great arteries (type I) depends on the balance between systemic and pulmonary blood flow (PBF). At birth, PBF may be antegrade through the right ventricular outflow tract, retrograde from a patent ductus arteriosus (PDA), or both. Changes after birth may upset the balance of systemic and PBF, including falling pulmonary vascular resistance, spontaneous restriction or closure of the PDA, and increasing right ventricular outflow tract obstruction. These changes require appropriately timed surgical procedures to adjust PBF. Procedures aimed at augmenting PBF include systemic to pulmonary artery shunt (SPS) and superior cavopulmonary connection (SCPC); those aimed at reducing PBF are interruption of the PDA, pulmonary artery banding (PAB), and interruption of the main pulmonary artery (MPA).

When PBF has $\geq$2 sources, either present or potential, that are either naturally occurring or secondary to intervention, management of competing PBF is controversial.[1-4] The Congenital Heart Surgeons' Society (CHSS) sought to determine the importance of surgical options in managing competing sources of PBF in a multicenter inception cohort of patients with tricuspid atresia type I, by characterizing the association between management of PBF at the initial and/or subsequent staged procedures and survival to a Fontan/Kreutzer (Fontan) operation.

METHODS

Study Cohort and Data Acquisition

Between January 1999 and October 2013, a total of 303 infants diagnosed with tricuspid atresia type I at <3 months of age were prospectively enrolled from 1 of 34 CHSS member institutions (Table E1). Tricuspid atresia type I was defined as failure of the tricuspid valve to develop, preventing a direct communication between the right atrium and the right ventricle, with concordant ventriculo-arterial connection. Further classification of the diagnosis was made based on status of the right ventricular outflow tract morphology at the time of diagnosis, according to institution echocardiogram reports, and included (Figure E1): 51 patients (17%) with pulmonary valve atresia (type Ia); 157 (52%) with restricted right ventricular outflow tract (type Ib); and 95 (31%) with an unrestricted right ventricular outflow tract (type Ic). Initial (index) intervention at a non-CHSS institution precluded patient enrollment. All treatment strategies were undertaken at the discretion of treating physicians.

Institutional participation and submission of patient information was confidential and voluntary. The CHSS Data Center and all participating institutions obtained ethics board approval, and parental consent was obtained before enrollment. Demographic, morphologic, and procedural data were abstracted from institutional medical records, including echocardiogram, surgical, and interventional reports. Baseline morphologic characteristics were defined based on initial echocardiogram reports. Subsequent morphologic details were abstracted from the echocardiogram reports that most immediately preceded the corresponding surgical intervention. Table E2 summarizes morphologic and demographic characteristics for all 303 patients.

Statistical Analysis

The goals of this analysis included: describing patient characteristics and management strategies for competing sources of PBF at the initial procedure and at subsequent staged operations, and to relate these as incremental risk factors to the time-related freedom from death. We used competing-risks methodology to account for the difference in timing associated with surgical strategies.[5,6]

Competing-risks analysis. Competing-risks analyses were used to examine the rates of transition from an initial state to mutually exclusive, time-related end-states (hazard function) of various procedure types or death without a corresponding procedure.[7] Given that the initial surgical strategy is largely dictated by the relative anatomic and physiologic differences among patients with tricuspid atresia, competing-risks analyses were performed in a similar manner for each of the following mutually exclusive, competing outcomes: (1) From the date of diagnosis to either death or Fontan operation for all 303 patients. For this analysis, management strategies at staged operations (SPS, PAB, and SCPC) were considered time-dependent covariables. (2) An analysis was performed from the initial SPS operation (n = 189) to either death or staged SCPC; and (3) from the first SCPC operation (n = 277; either staged or primary) to death or Fontan operation.

For each competing-risks analysis, non–risk-adjusted nonparametric estimates for time-related freedom from all-cause mortality or the specified procedure type were plotted using the Kaplan-Meier method. The underlying hazard functions were modeled parametrically to determine multiple phases of risk, as previously described.[8] Subsequently, risk-hazard analyses were performed to identify risk factors associated with each model.

Risk-hazard analysis and presentation. The status of PBF before, and management of PBF during, each procedure; all transcatheter interventions; and subsequent unplanned surgical operations were analyzed as covariables. Selected measurements were related to body surface area and converted to z-scores on the basis of published normative data, if available.[9] Variables with excessive (>75%) missing values were excluded during multivariable analysis. To account for the remaining missing values, 5-fold multiple imputation was performed.[10] Final variable selection was guided by bootstrap bagging (n = 500, threshold for inclusion P = .1). Variables selected in at least approximately 50% of resamples or their clusters were considered reliable for inclusion. A P < .07 was considered significant for final variable retention.

Reported parameter estimates represent the contribution of a variable to the overall model. Continuous variables were compared among 3 groups with the Kruskal-Wallis test, using Wilcoxon rank-scores. The frequencies for categoric variables were compared using a χ^2 test of independence or the Fisher exact test, as appropriate. Data are presented as median with range, mean $\pm$ SD or frequency, with missing values indicated where appropriate. Data analyses were performed with SAS statistical software (version 9.2; SAS Institute, Inc, Cary, NC). Additional information regarding statistical techniques is given in Appendix 1.

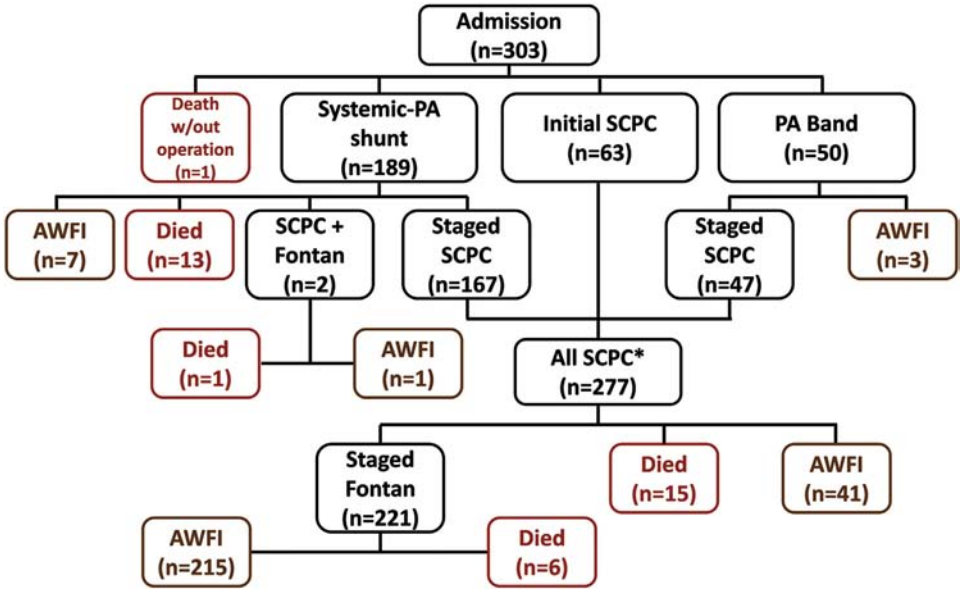

FIGURE 1. Flow chart depicting events from diagnosis for 303 children with tricuspid atresia type I. One patient died without any surgical intervention. All other children underwent initial palliation with systemic-PA shunt, PA band, or initial SCPC. After the index systemic-PA shunt, 13 children died; 167 went on to an interval SCPC; 7 are AWFI; and 2 had single-staged completion Fontan/SCPC, 1 of whom died. For children who had an index PA band, 47 went on to an interval SCPC, and 3 are AWFI. From the initial 303 children, 277 transitioned to an all SCPC. After SCPC, 15 children died before Fontan completion, 41 are AWFI (median age: 1.3 years; range: 0.1-12.4 years), and 221 achieved a staged Fontan operation (median age at Fontan: 2.8 years; range: 0.9-7.6 years). A total of 7 deaths occurred after Fontan operations (1 after single-staged Fontan/SCPC), and 215 patients are AWFI. *PA*, Pulmonary artery; *SCPC*, superior cavopulmonary connection; *AWFI*, alive without further intervention. *"All SCPC" consists of patients who have achieved SCPC after initial systemic-PA shunt, PA band, or had no previous surgical intervention.

RESULTS

Surgical Management

Initial surgical management (Figure 1) for 302 patients included SPS (n = 189; 62%), PAB (n = 50; 17%), or SCPC (n = 63; 21%), with 1 death before any surgical intervention (Table E3). The SPS procedures included 183 (97%) modified Blalock-Taussig shunts (originating from an aortic arch branch), and 6 (3%) central shunts originating from the aorta. For 25 patients, a concomitant MPA intervention (21 with MPA closure and 4 with MPA banding) was performed during initial SPS. MPA closure and banding were considered jointly as MPA interruption. Of 189 SPS procedures, 49 (26%) were performed using cardiopulmonary bypass.

Superior cavopulmonary connection was undertaken in 277 patients, as either a primary or staged operation. Of these 277 patients, 167 (60%) had an index SPS, and 47 (17%) had an index PAB. For 63 (23%) patients, SCPC was the first operation. Concomitant pulmonary arterioplasty during SCPC, on either the right, left, or both branch pulmonary arteries was performed more frequently in patients who underwent an initial SPS (40%), compared with those who had an index PAB (30%) or SCPC (17%; chi-square $P = .004$). Of 277 SCPC operations, 237 (86%) were performed using cardiopulmonary bypass. Table 1 summarizes the source of native and accessory PBF and management decisions for augmentation and/or restriction of PBF at each staged surgical procedure.

Mortality

Of the 36 total deaths (Table E4), 29 occurred before a Fontan operation. Of these 29 deaths, 1 child died without intervention; 23 deaths occurred in patients who underwent an initial SPS; 2 deaths occurred in patients who underwent an initial PAB; and 3 deaths occurred in patients who had an SCPC without a previous surgical operation.

Overall Survival Before Fontan Operation

For all patients, the unadjusted 6-year survival before Fontan was 88% ± 2% (n = 303; Figure 2, *A*). Patients who underwent an initial SPS had significantly lower 6-year, risk-adjusted survival (85% ± 5%; $P = .041$) relative to all other patients (93% ± 3%; Figure 2, *B*). Among all patients, an increased risk of death before a Fontan operation was incurred if SPS was performed with a concomitant MPA interruption. For these patients, the hazard (instantaneous risk) for death was increased early, and it remained elevated relative to patients who had an SPS without MPA interruption (Figure 3, *A*). Additional factors associated with an increased risk of death for all patients included lower birth weight ($P = .046$); and more-than-mild mitral valve regurgitation on baseline echocardiogram trended

TABLE 1. Procedural characteristics and status of competing PBF

Variable	Value	Deaths before Fontan (after SCPC)
Initial SPS	189	23 (10)
Indexed shunt size (mm/kg), mean ± SD	1.3 ± 0.34	—
Performed on CPB	49	4
Transcatheter procedure before initial SPS		
Atrial septostomy before SPS	20	1
Ductal stent	1	0
Shunt origin		
Innominate artery	133	7
Subclavian artery	39	5
Carotid artery/other	7	1
Aorta	7	1
Shunt connection		
Right pulmonary artery	148	16 (6)
Pulmonary artery, left or branch, bifurcation	21	4 (2)
MPA	20	3 (2)
Persistent PDA after SPS	7	4
Status of MPA after SPS		
Pulmonary valve atresia	51	5 (2)
No MPA intervention	113	13 (4)
MPA intervention	25	5 (4)
MPA closed	21	4
MPA banding	4	1
SCPC after SPS	167	10
Hemi-Fontan	28	2
BDCPA	139	8
Performed on CPB	148	8
MPA status after SCPC		
Pulmonary valve atresia	45	2
Uninterrupted MPA	50	2
Previously banded MPA/intact after SCPC	3	0
Previously banded MPA/interrupted at SCPC	1	1
MPA closed at SCPC	48	2
MPA closed before SCPC	20	3
Bilateral branch pulmonary artery plasty at SCPC	26	5
Initial PAB	50	2 (after SCPC)
Atrial septostomy before PAB	5	0
PDA closed after PAB	50	2
SCPC after PAB	47	2
Hemi-Fontan	7	0
BDCPA	40	2
Performed on CPB	43	2
MPA status after SCPC		
MPA interrupted at SCPC	43	2
MPA left intact at SCPC	4	0
Bilateral branch pulmonary artery plasty at SCPC	8	0
SCPC as initial procedure	63	3
Atrial septostomy before SCPC	8	0
Hemi-Fontan	7	0
BDCPA	56	3
Performed on CPB	56	3
MPA status after SCPC		
MPA interrupted at SCPC	44	3
MPA left intact at SCPC	19	0
Bilateral branch pulmonary artery plasty	2	0

(Continued)

TABLE 1. Continued

Values are n, unless otherwise indicated. Shown in the table are the procedural characteristics and status of competing PBF at the time of the initial procedure and at subsequent SCPC for: patients who underwent an initial SPS (n = 189); patients who underwent an initial PAB (n = 50); and patients who had an initial SCPC (n = 63) as the primary procedure. *Ten of the 23 deaths in the SPS group occurred after SCPC. *PBF*, Pulmonary blood flow; *SPS*, systemic to pulmonary artery shunt; *SD*, standard deviation; *SCPC*, superior cavopulmonary connection; *CPB*, cardiopulmonary bypass; *PDA*, patent ductus arteriosus; *MPA*, main pulmonary artery; *BDCPA*, bidirectional cavopulmonary anastomosis; *PAB*, pulmonary artery band.

toward significance ($P = .067$; Table 2). Use, or lack of use, of extracorporeal circulation, at SPS ($P = .78$) or at subsequent SCPC ($P = .70$) did not significantly influence the risk of death.

Transition to Superior Cavopulmonary Connection: After Systemic to Pulmonary Artery Shunt

For patients who had an initial SPS or PAB, all deaths before SCPC (n = 13) occurred after SPS. For survival before SCPC for 189 patients who underwent SPS, the estimated 1-year, risk-adjusted survival was 93% ± 2%. An increased risk of death after SPS was associated with a lower immediate postoperative oxygen saturation, recorded from pulse-oximetry (SpO_2) before extubation (parameter estimates = −0.15, $P < .001$). The association with poor survival seems most predominant at SpO_2 levels <75%-80% (Figure 4). Additionally associated with poor survival was a persistent PDA (ie, did not close spontaneously before SPS or was not occluded during SPS; n = 7). The 1-year survival after SPS was 76% when the PDA remained open, versus 97% when the PDA spontaneously closed before SPS, or was occluded during SPS (parameter estimates = 2.5, $P < .001$). Seven unplanned surgical reoperations occurred after SPS (5 procedures for shunt thrombosis and 2 PDA closures). Unplanned reoperations were not associated with an increased risk of death.

Transition to Superior Cavopulmonary Connection: After Pulmonary Artery Banding

For 50 patients who underwent an initial PAB, no deaths occurred before SCPC; 47 transitioned to SCPC; and 3 were alive, awaiting further operation. Thus, these patients had no associated risk factors for death before SCPC. The PDA closed spontaneously, before PAB, or was occluded during PAB, for all patients.

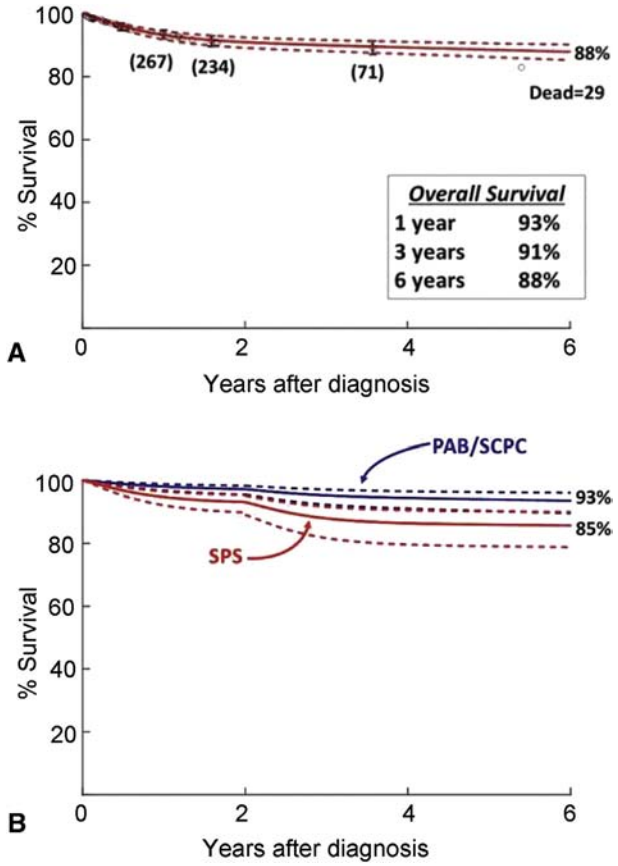

FIGURE 2. A, Overall unadjusted, 6-year survival without Fontan for all 303 study patients. The hazard (instantaneous risk) for death is characterized by an early phase (23 events), with a rapid decline before 2 years, followed by a late low constant phase (6 events). Survival was 93% ± 1%, 91% ± 1%, and 88% ± 2%, at 1, 3, and 6 years, respectively. *Circles* represent nonparametric estimates at events (deaths); *numbers in parentheses* represent the number of patients at risk at that point; s*olid lines* represent parametric determinants of continuous point estimates. *Dashed lines* and *bars* enclose 68% confidence intervals. B, Overall estimated, risk-adjusted, 6-year survival without Fontan, stratified by initial procedure. Each curve represents the predicted survival with all risk factors held constant at the median value, except for the initial procedure. The transition (break) in the curve occurs at 0.7 years (mean time of SCPC) and accounts for the time-varying change in survival that occurs with SCPC completion. *The blue curve* represents risk-adjusted predicted survival in 113 patients who underwent either an initial PAB or SCPC (93% ± 3%); *the red curve* represents the risk-adjusted predicted survival for 189 children who underwent an initial SPS procedure (85% ± 5%). S*olid lines* represent parametric determinants of continuous point estimates, and *dashed lines* and *bars* enclose 68% confidence intervals. Median/median value for risk factors: birth weight: 3.1 kg; >mild mitral valve regurgitation: 0.28; and interrupted main pulmonary artery at initial SPS: 0.09. *SPS*, Systemic to pulmonary artery shunt; *PAB*, pulmonary artery band; *SCPC*, superior cavopulmonary connection.

Outcomes After Superior Cavopulmonary Connection for all Patients

For 277 patients who transitioned to SCPC, the unadjusted 4-year survival before Fontan was 93% ± 2%. Of the 15 deaths after SCPC, 10 occurred in patients who had an initial SPS; 2 patients had a PAB; and 3 patients had no surgical procedure before SCPC (Figure E2). Risk-adjusted analysis demonstrated an increased risk of death for patients who had MPA intervention (closure n = 20, with 3 deaths; banding n = 4, with 1 death) at the time of initial SPS (parameter estimate = 1.5, $P = .020$). When the risk-adjusted hazard for death after SCPC was stratified based on initial procedure (SPS, PAB, or SCPC), the resulting hazard functions were nearly equivalent, except for patients who had concomitant MPA intervention at SPS (Figure 3, *B*). These patients had a sustained, increased risk for death and overall lower 4-year risk-adjusted survival (78%), compared with those who had an initial SPS without MPA intervention (94%) or those who had an initial PAB or SCPC (95%).

These results are consistent with the findings of the overall analysis for all 303 patients. The implication is that MPA intervention at initial SPS is associated with an increased risk of death that persists beyond SCPC. Based on univariable analysis, bilateral branch pulmonary artery arterioplasty at the time of SCPC was a risk factor for death before Fontan (parameter estimate = 1.3; $P = .017$), irrespective of PBF status through the MPA. However, on multivariable analysis, branch pulmonary artery arterioplasty did not remain a significant risk factor for death.

At the time of SCPC, 212 (77%) of 277 patients had antegrade PBF through the MPA—only 65 patients who underwent an initial SPS had no antegrade PBF at SCPC, secondary to either pulmonary valve atresia (n = 45) or MPA closure at the time of initial SPS (n = 20). For the 212 patients with antegrade PBF through the MPA at SCPC, closure was performed for 136 patients (49 SPS, 43 PAB, and 44 SCPC). Therefore, 76 patients had persistent antegrade PBF through the MPA, after SCPC. Eight deaths occurred among patients who underwent MPA closure during SCPC, versus only 2 deaths among those in whom the MPA was uninterrupted. However, MPA interruption at SCPC was not a significant risk factor for death (parameter estimate = −1.0; $P = .18$).

Excluding the167 patients who underwent SCPC after initial SPS, 110 patients had antegrade PBF at SCPC, including: 47 patients who had a stage-1 PAB; and 63 for whom SCPC was the primary operation. For these patients, 87 underwent MPA interruption (5 deaths before Fontan completion), and 23 had an MPA left intact (0 deaths). Although these results are suggestive of a trend toward worse survival for patients without antegrade PBF after SCPC, MPA closure was not a significant risk factor for death in this subset analysis (log-rank test, $P = .22$).

Finally, for 256 patients with echocardiogram reports after SCPC, 9 had moderate or severe systolic right ventricular dysfunction, and of these, only 1 had persistent antegrade PBF; and 7 of the 256 had moderate or severe mitral valve regurgitation.

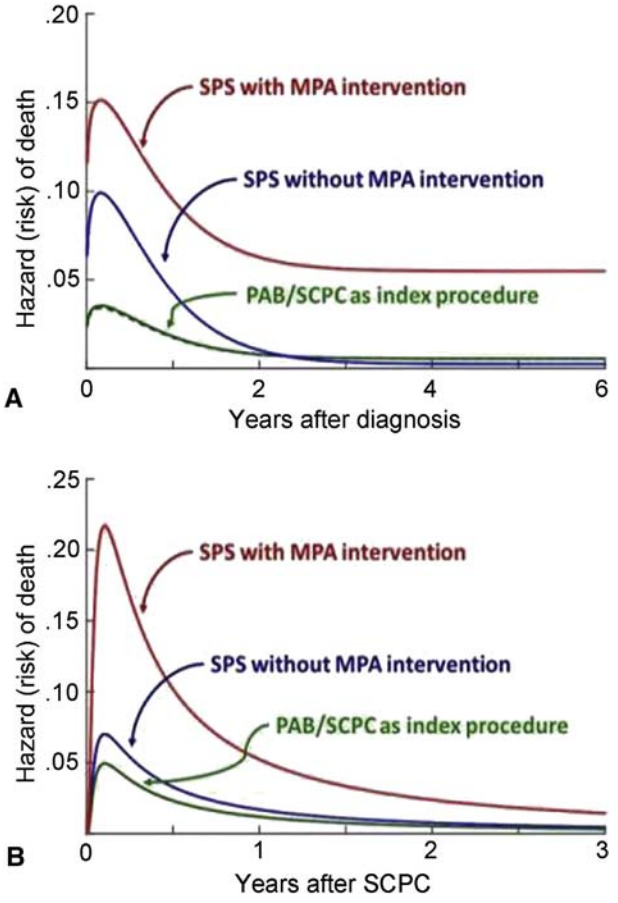

FIGURE 3. A, Overall estimated hazard (instantaneous risk) for death after diagnosis, for all 303 children diagnosed with tricuspid atresia. *The red curve* represents 25 patients who underwent an initial SPS with concomitant MPA intervention (closure = 21, band = 4). *The blue curve* represents 164 patients who underwent an initial SPS procedure without MPA intervention. The *green curve* represents all other patients, including 50 who had an initial PAB, and 63 who had an SCPC without any previous intervention. B, The estimated hazard (instantaneous risk) for death after SCPC diagnosis for 277 children who underwent SCPC. The *red curve* represents 24 children who underwent an initial SPS with concomitant MPA intervention and subsequently transitioned to SCPC. The *blue curve* represents 143 children who underwent an initial SPS procedure without MPA intervention and subsequently transitioned to SCPC. The *green curve* represents all other children, including 47 who had an initial PAB, and 63 who had an SCPC without any previous intervention. The early risk of death is elevated in all patients undergoing initial SPS, compared with all other patients. This risk decreases and becomes nearly equivalent for all children after transition to SCPC, except for children who underwent concomitant MPA intervention at the time of SPS. For these children, the elevated risk of death persists beyond stage-2 SCPC. *SPS*, Systemic to pulmonary artery shunt; *MPA*, main pulmonary artery; *PAB*, pulmonary artery band; *SCPC*, superior cavopulmonary connection.

DISCUSSION

Management of competing sources of PBF at initial palliation and subsequent staged operations remains a surgical dilemma for patients with tricuspid atresia type I.[1-4] Although anatomic and physiologic characteristics may necessitate a stage-1 SPS operation, our multi-institutional review demonstrated that SPS is an independent risk factor for death. Factors potentially mitigating the risk of death after SPS include spontaneous or surgical PDA closure and avoidance of MPA intervention at the time of SPS.

Outcomes After Systemic to Pulmonary Artery Shunt

The finding that SPS operations are associated with increased mortality is consistent with previous studies identifying Blalock-Taussig shunt procedures as a risk factor for death in patients with univentricular hearts.[11,12] Systemic to pulmonary artery shunts are, themselves, associated with acute complications that contribute to patient morbidity and mortality.[13] Alternatively, the need for SPS is sometimes a surrogate for disease complexity or perioperative morbidity, including: noncardiac anomalies, low birth weight, preoperative mechanical support, and acidosis or circulatory shock.[11,12] Although the data necessary to determine preoperative clinical status were limited, low birth weight was identified as a significant risk factor for death. This finding is consistent with previous reports identifying low birth weight as a strong and reliable predictor of poor survival for neonates with congenital heart lesions.[14] Additionally, the use of extracorporeal circulation may have been performed in a high-risk subset of patients undergoing SPS; however, we did not find this factor to have a significant effect on survival.

Our results imply that lower post-SPS oxygen saturations are associated with an increased risk of death. Similarly, a lower oxygen saturations may be a proxy for decreased cardiac output and associated preoperative morbidity. Alternatively, the results may suggest that an imperfect PBF-to-systemic-blood-flow ratio early after SPS coincides with poor survival. However, in the setting of mechanical ventilation, oxygen saturations are artificially manipulated, and pulse oximetry measurements are often variable. Although we considered various forms of risk stratification based on shunt size, shunt origin, and birth weight, the variability of blood oxygen level measurements cannot be completely avoided. Interpretation of this finding may not necessarily suggest that inflating postoperative oxygen saturations will improve survival, but rather that lower saturations are a marker for poor outcomes.

Management of accessory pulmonary blood flow after systemic to pulmonary artery shunt. Despite the risks associated with SPS operations, augmentation of PBF via SPS is the only viable treatment option for some patients, which should be considered when discussing factors that potentially mitigate the risk of death after SPS operations. Given this caveat, our study suggests that a PDA that spontaneously closes before the SPS procedure, or is surgically

TABLE 2. Risk factors associated with death before Fontan for all 303 children

Risk factors	Parameter estimate ± SD	*P* value	Reliability (%)
Early phase			
Intercept	−4.8 ± 0.85	<.001	—
SPS as initial procedure	1.2 ± 0.58	.041	43
Birth weight*	3.2 ± 1.6	.046	57
MV regurgitation on baseline echocardiogram	0.8 ± 0.44	.067	47
Constant phase			
Intercept	−5.5 ± 1.1	<.001	—
SPS as initial procedure†	−0.8 ± 1.5	.59	—
MPA intervention at initial SPS procedure	3.4 ± 1.5	.021	48

Incremental risk factors associated with death before Fontan for all 303 children with tricuspid atresia after diagnosis, with associated parameter estimates ± SD, *P* values, and bootstrap relatability (n = 500). The rate of transition (hazard function) was modeled from the date of diagnosis (time-zero) to death before Fontan or Fontan operation. *SD*, Standard deviation; *SPS*, systemic to pulmonary artery shunt procedure; *MV*, mitral valve; *MPA*, main pulmonary artery. *Birth weight was included as inverse transformation. †SPS was included in the constant phase as an interaction term, to ensure that MPA intervention was not significant because of the corelationship with SPS.

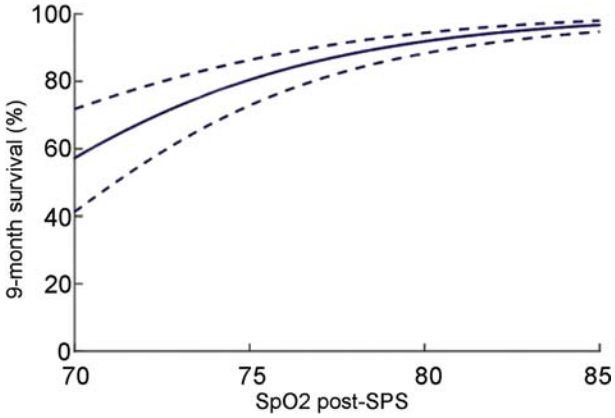

FIGURE 4. The 9-month estimated survival without achievement of Fontan after SPS, stratified by postprocedure SpO2 levels. The SpO2 levels were recorded immediately postprocedure, via pulse oximetry, with patients still intubated. The associated risk of death is significantly increased ($P = .02$), as SpO2 levels decrease, and is most predominate at SpO2 levels of <75% to 80%. The *solid line* represents continuous parametric estimates; *dashed lines* indicate 68% confidence intervals. *SpO2*, Oxygen saturation; *SPS*, systemic to pulmonary artery shunt.

closed during the procedure, avoiding MPA interruption at the time of the SPS, is associated with a decreased risk of early mortality.

Current evidence supporting PDA closure for patients undergoing SPS operations is mixed.[12,15,16] Those in favor of a persistent PDA after an SPS procedure suggest that it improves outcomes in the setting of acute shunt thrombosis.[16] However, others suggest that a PDA: (1) potentiates shunt thrombosis via a steal phenomenon and is associated with poor outcomes[15]; or (2) does not have a significant association with the time-related risk of death.[12]

Limiting MPA disruption at the time of SPS was associated with better long-term survival in our series. Potential explanations of this result are related to the findings from previous analyses describing the adverse effects of shunt physiology on PBF. Prosthetic shunt physiology alters, or with patient growth, may impede PBF, resulting in an inability to sustain pulmonary artery development. The restricted PBF associated with shunt physiology may contribute to abnormal pulmonary vasculature and distorted branch pulmonary arteries at the time of SCPC.[2,13] Furthermore, previous reports have found associations with shunt procedures and the requirement for pulmonary arterioplasty during the SCPC procedure.[11] Branch pulmonary artery arterioplasty was not an independent risk factor for death in our series; however, patients who underwent an SPS had an increased incidence of branch pulmonary artery arterioplasty at the time of SCPC, compared with patients who had an initial PAB or SCPC. This was especially true for those patients who had interruption of the MPA at the time of SPS, suggesting that branch pulmonary artery arterioplasty may be a corollary for MPA interruption during SPS procedures.

Antegrade Pulmonary Blood Flow Through the Main Pulmonary Artery

Although our results suggest an association between poor survival and MPA intervention at the time of SPS, this does not necessarily implicate antegrade PBF as a risk factor. Another possibility is that overmanipulation of the MPA early on can have longer-term repercussions. Furthermore, we found no association (either positive or negative) between persistent antegrade PBF through the MPA and the risk of death after SCPC.

A recent report, comparing 57 children with antegrade PBF (group 1) and 54 without antegrade PBF (group 2) after SCPC, reported 1 and 6 deaths before a Fontan operation for groups 1 and 2, respectively. This difference resulted in significantly better 5-year survival for children in group 1 with antegrade PBF[17] (log-rank test; $P = .03$). In our series, for 110 children in whom the initial procedure was a PAB or SCPC, all 5 deaths occurred in children who underwent MPA closure during the SCPC procedure. Although this did not result in a statistically significant difference in survival in our series, continued follow-up and increased enrollment may show that MPA closure at SCPC is an important predictor of survival.

Evidence continues to mount for the potential benefits of maintained forward flow through the MPA after stage-2 SCPC. Such benefits include: improved oxygenation, continued pulmonary artery growth, protection of normal pulmonary vasculature with decreased pulmonary vascular resistance, and prevention of pulmonary arteriovenous malformations.[17-20] In addition to improving early outcomes after SCPC, these benefits may have an extended effect of delaying time to failure after Fontan operations.

Drawbacks of the Fontan circulation include an increase in pulmonary vascular resistance with elevated systemic venous pressures and a chronic low-output state, eventually leading to progressive ventricular dysfunction and failure of single-ventricle palliation.[19-21] Additionally, the increased venous pressure negatively affects hepatic tissue and function.[22] Therefore, postponing Fontan palliation, by maintaining antegrade PBF and extending the time to Fontan operation, may theoretically result in a longer time before Fontan failure, a reduced negative impact on hepatic function, and potentially improved survival after Fontan palliation.[21-23]

However, the potential benefits of persistent PBF must be weighed against the associated risks. Adverse effects of increased PBF are related to greater systemic venous pressure, which has been associated with longer time to extubation, excessive pleural effusion, and increased time to discharge.[17,24] Furthermore, the increased volume load on a single ventricle may contribute to reduced ventricular and atrioventricular valve performance.[25] In addition, for children with tricuspid atresia type Ia, accessory sources of PBF have not been associated with improved outcomes and are therefore not a viable option.[2,12,15]

Study Limitations and Future Analyses

The primary limitations of this study were related to the lack of quantitative morphologic and physiologic data. Interstage longitudinal measurements on pulmonary artery size, ventricular dimensions and function, oxygen saturation, and pulmonary artery pressure were limited. These measurements would have helped to determine the influence of various management strategies in the setting of relatively few events (deaths). Specifically, an expanded data set may have helped identify how various surgical strategies influenced the growth of anatomic structures and single-ventricle performance. The analysis was additionally limited by a lack of comprehensive data regarding surgical details, clinical status, and causes for death. Although occasionally documented in clinical records, this information was not consistently available for each patient. Finally, the analysis was subject to the limitations on any prospectively gathered observational studies. Specifically, the intent of clinical decision making could not be inferred from medical records.

The current study is an important extension to the previous CHSS study using a similar cohort.[26] Our current analysis is enhanced by longer follow-up, a study population with twice as many patients, and contemporary statistical methodologies. Potential areas of focus for future analyses include a more thorough examination of how MPA intervention influences pulmonary artery growth and exploring outcomes after Fontan palliation. This evaluation may include a quantitative measure of the impact of the MPA closure technique at or before Fontan palliation on post–Fontan surgery complications.

CONCLUSIONS

Initial patient morphology and evolving physiology will predicate management strategies for accessory PBF. For many patients, augmentation of PBF via a systemic to pulmonary artery shunt may be the only viable option. However, SPS itself confers a higher-risk status for these patients. Avoidance of native MPA intervention during initial SPS, and closure of a PDA, are associated with improved survival. Furthermore, native MPA intervention—or lack thereof—during SCPC procedures did not significantly affect survival. Among all patients, we did not identify an association between antegrade PBF through the MPA and mortality. Ultimately, each clinical situation is unique, and management of accessory PBF must be made by the operating surgeon.

Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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Key Words: tricuspid atresia, accessory pulmonary blood flow, systemic to pulmonary artery shunt, congenital heart disease

APPENDIX 1: STATISTICAL METHODS

Flow charts were created to track patients through multiple consecutive procedures and death. Competing-risks analyses were used to examine rates of transition from an initial state (hazard function) to the mutually exclusive time-related events of various procedures, and death without that procedure. Procedures included as end-states were the initial procedure (SCPC was the initial procedure for 63 patients), subsequent SCPC, and Fontan operation. This information was used to determine the proportion of patients who reached these events or states at any given time after the initial state.

Competing-risks analyses were performed in a similar manner for each of the following: (1) from initial admission to a CHSS member institution, to either death before Fontan operation, or completion of Fontan procedure; (2) from an initial SPS procedure to SCPC, or death before SCPC; and (3) from SCPC to either death before Fontan operation, or subsequent transition to Fontan operation. Initially, a competing-risks hazard model for all 303 patients was created, starting from the date of diagnosis. Date of diagnosis provided a uniform start date (time zero) and ensured that our survival analysis incorporated 100% of eligible patients.[1,2]

A unique feature of this model was the incorporation of both time-independent (baseline morphologic and demographic) variables, as well as time-dependent variables, such as major surgical operations (SPS, PAB, SCPC). The motivation for the subsequent competing-risks model for patients who had an initial SPS (n = 189) was driven by 2 primary factors: (1) death before SCPC occurred exclusively in patients who had a primary SPS procedure (with the exception of 1 death before any intervention); and (2) patients undergoing SPS were found to have an increased risk of death before Fontan, based on the initial competing-risks model. A competing-risks model was created with the subset of patients who achieved SCPC (n = 277), to evaluate how management of PBF at SCPC influences death before Fontan palliation.

Informative imputation (multiple imputation) based on available data was used to replace missing values. Relevant missing value indicator variables were created and included in multivariable analyses to adjust for possible bias introduced by missing data. Missing value indicator variables were entered into final multivariable models as appropriate. For continuous variables, various mathematical transformations were tested for optimal calibration of the relationship to risk, and the significance of various interaction terms was explored. The multivariable models were solved in a competing-risks format for multiple combinations of risk factors, to explore the effect magnitude of these factors and predict outcomes for patients with given characteristics.

E-References

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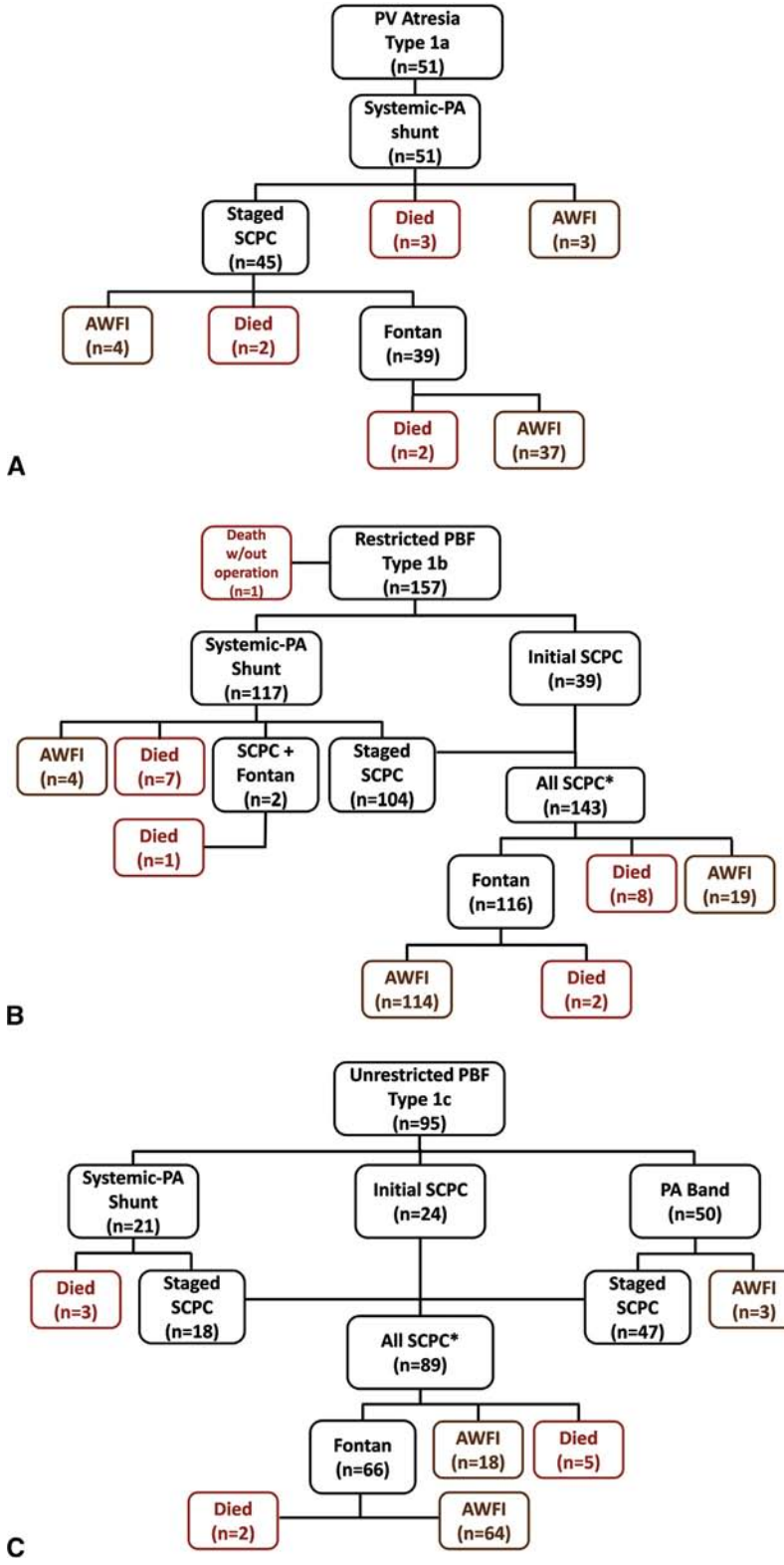

FIGURE E1. Flow chart depicting events from diagnosis for 303 children, based on morphology of the RVOT at diagnosis, according to institution echocardiogram reports for: (A) all 51 children with pulmonary valve atresia (type Ia); (B) 157 children with restricted RVOT PBF (type Ib); and (C) 95 children with unrestricted RVOT PBF (type Ic). Systemic-PA shunt indicates children who had an initial systemic to pulmonary artery shunt operation. PA band indicates children who had an initial PA banding operation. Initial SCPC indicates children who had this operation without a previous surgical operation. *PV*, Pulmonary valve; *PA*, pulmonary artery; *SCPC*, superior cavopulmonary anastomosis; *AWFI*, alive without further intervention; *PBF*, pulmonary blood flow. *All SCPC consists of patients who have achieved SCPC after initial systemic-PA shunt or PA band, or had no previous surgical operation.

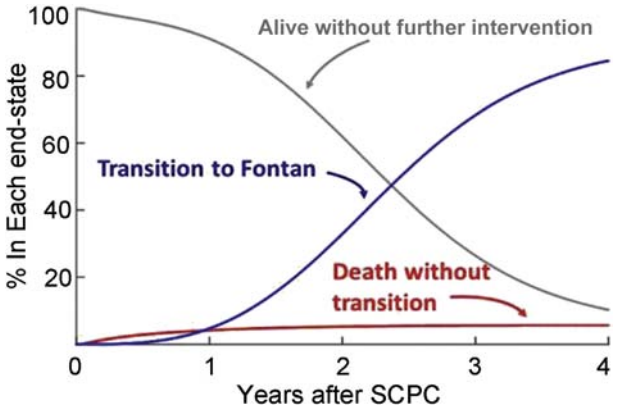

FIGURE E2. Competing end-states depiction of events following SCPC for 277 children with tricuspid atresia. All children were alive at the time of procedure and thereafter entered 1 of 3 mutually exclusive end-states: (1) alive without further intervention (*gray curve*); (2) transition to Fontan (*blue curve*); (3) death (*red curve*). At 4 years after SCPC, 10% of children were alive without further intervention, 83% had transitioned to Fontan, and 7% had died. Independent of time, 221 children underwent a Fontan operation; 15 deaths occurred without transition to Fontan palliation. Not represented are the 6 deaths that occurred after Fontan operation. At any point in time, the sum of the proportions of children in each state must be 100%. *Solid lines* represent continuous parametric estimates. *SCPC*, Superior cavopulmonary anastomosis.

TABLE E1. Participating Congenital Heart Surgeons' Society member institutions

Alfred I. duPont Hospital for Children, Wilmington, Del
All Children's Hospital, St Petersburg, Fla
Ann & Robert H. Lurie Children's Hospital of Chicago, Chicago, Ill
Benioff Children's Hospital, University of California at San Francisco, San Francisco, Calif
Boston Children's Hospital, Boston, Mass
Cardinal Glennon Children's Medical Center, St Louis, Mo
Children's Hospital of Los Angeles, Los Angeles, Calif
Children's Hospital of Michigan, Detroit, Mich
Children's Hospital of Pittsburg, Pittsburg, Pa
Children's Mercy Hospitals and Clinics, Kansas City, Mo
Children's National Heart Institute, Children's National Medical Center, Washington, DC
Christ Hospital Medical Center, Oak Lawn, Ill
Cincinnati Children's Hospital, Cincinnati, Ohio
Comer Children's University of Chicago Medical Center, Chicago, Ill
C. S. Mott Children's Hospital, Ann Arbor, Mich
Emory Clinic, Atlanta, Ga
Loma Linda University Health Care, Loma Linda, Calif
Medical University of South Carolina, Charleston, SC
Montreal Children's Hospital, Montreal, Quebec, Canada
Monroe Carell Jr Children's Hospital at Vanderbilt, Nashville, Tenn
Oregon Health & Science University, Portland, Ore
Penn State Milton S. Hershey Medical Center, Hershey, Pa
Primary Children's Medical Center, Salt Lake City, Utah
Rady Children's Hospital, San Diego, Calif
Riley Hospital for Children, Indianapolis, Ind
St. Louis Children's Hospital, St Louis, Mo
St. Christopher's Hospital for Children, Philadelphia, Pa
Stollery Children's Hospital Foundation at University of Alberta, Alberta, Edmonton, Canada
The Children's Heart Clinic of Minnesota, Minneapolis, Minn
The Children's Hospital at Denver, Denver, Colo
The Hospital for Sick Children, Toronto, Ontario, Canada
University of California at Los Angeles, Los Angeles, Calif
University of Iowa Children's Hospital, Iowa City, Iowa

TABLE E2. Initial demographic and morphologic characteristics for all 303 children with tricuspid atresia

Variable	Value	Deaths before Fontan (n = 29)	Missing
Demographic characteristics			
Birth weight (kg)	3.2 (1.0-4.7)	—	14
BSA (m^2)	0.21 (0.1-0.28)	—	14
Age at admission (d)	0 (0-620)	—	0
Male; female	153; 150	17; 12	0
Morphologic characteristics	—	—	—
Pulmonary atresia, type Ia	51 (17)	5 (2)	0
Restricted PBF, type Ib	157 (52)	16 (5)	0
Unrestricted PBF, type Ic	95 (31)	8 (3)	0
Aortic valve stenosis	9 (3)	2 (1)	14
Aortic valve regurgitation	22 (7)	4 (1)	30
Mitral valve regurgitation	70 (23)	10 (3)	44
Left ventricle dysfunction	11 (4)	0 (0)	17
Restrictive ASD	24 (8)	1 (1)	37
Restrictive VSD	108 (36)	7 (2)	13
Aberrant SVC	28 (9)	2 (1)	70
Pulmonary valve annulus (cm)	0.64 ± 0.20	—	117
Right branch PA z-score	−1 (−7 to 4)	—	115
Left branch PA z-score	−1 (−8 to 4)	—	120
Minimum branch PA z-score*	−2 (−10 to 4)	—	115
Initial admission saturation (%)	83.4 ± 10.1	—	140
Initial discharge saturation (%)	86.0 ± 5.7	—	172
Post-SPS saturation (%)	85 ± 6	—	44

Values are n, n (%), median and range, or mean ± SD. Morphology of the right ventricular outflow tract was based on institution echocardiogram report. *BSA*, Body surface area; *PBF*, pulmonary blood flow; *ASD*, atrial septal defect; *VSD*, ventricular septal defect; *SVC*, superior vena cava; *PA*, pulmonary artery; *SPS*, systemic to pulmonary artery shunt. *Minimum z-score measurement of either the right or left PA branch.

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TABLE E3. Patient characteristics and outcomes, stratified by initial surgical procedure, for 302 children who underwent SPS, PAB, or SCPC

Variable	SPS (n = 189)	PAB (n = 50)	SCPC (n = 63)	*P* value
Age at initial procedure (d)	26 ± 29	33 ± 29	164 ± 88	<.01
Birth weight (kg)	3.1 ± 0.65	3.2 ± 0.6	3.12 ± 0.65	.65
O_2 saturation on admission	83 ± 11	89 ± 8	85 ± 9	<.01
Left branch PA z-score	−1.5 ± 1.7	−0.3 ± 2.1	−0.6 ± 2	<.01
Right branch PA z-score	−1.7 ± 1.9	0 ± 2.2	−1.01 ± 1.77	.04
Minimum branch PA z-score*	−2 ± 2.7	−0.7 ± 2.1	−0.8 ± 2	.01
Restrictive VSD	79 (42)	3 (6)	25 (40)	<.01
Total deaths	29 (15)	3 (6)	3 (5)	.03
Fontan operation	140 (74)	31 (62)	52 (83)	.05

Values are mean ± SD, or n (%), unless otherwise indicated. *P* values for continuous variables are based on the Kruskal-Wallis test, using Wilcoxon rank scores. The frequencies for categoric variables were compared using the Fisher exact test. Birth weight was not significantly different among the 3 groups but is included for reference. *SPS*, Systemic to pulmonary artery shunt; *PAB*, pulmonary artery band; *SCPC*, superior cavopulmonary connection; O_2, oxygen; *PA*, pulmonary artery; *VSD*, ventricular septal defect. *Minimum z-score measurement of either the right or left PA branch.

TABLE E4. Details of all 36 deaths after diagnosis

RVOT	Initial operation (age [d])	Age of death (y)	PDA status after first procedure	PBF status after SCPC	Details of MPA closure	Complications/morbidities	Details related to death
1b	SPS (1)	0.5	Closed	Death before SCPC	n/a	Portal vein thrombosis 2/2 to umbilical catheter; Rx heparin and aspirin	Sudden circulatory compromise
1b	SPS (9)	1.6	Closed	Death before SCPC	n/a	—	Unknown cause of death at secondary institution
1b*	SPS (4)	2.9	Closed	− Antegrade PBF	MPA ligated at SCPC	—	Death after Fontan found on death registry, unspecified cause of death
1a	SPS (2)	1.1	Open, PGE discontinued	Death before SCPC/ pulmonary atresia	n/a	Return to catheter lab POD 2 for PDA closure, 2/2 to persistent PDA	Unspecified cause of death
1c	SPS (26)	3.6	Closed	− Antegrade PBF through MPA Bilateral BPA plasty at SCPC	PV oversewn at SPS	Mitral valve insufficiency after SCPC	Cardiac arrest requiring ECMO, MSOF
1b	SPS (49)	0.7	Closed	+ Antegrade PBF	MPA uninterrupted	Post-SCPC had poor O_2 saturation, return to OR for redo shunt	Decreased cardiac output with ultimate cardiac failure
1b	SPS (64)	1	Closed	− Antegrade PBF Bilateral BPA plasty at SCPC	PV oversewn at SCPC	—	Sudden death at home; no autopsy; unspecified cause of death
1b*	SPS (32)	0.95	Open, PGE discontinued	Jehovah's Witness: To avoid multiple operations, planned SCPC and Fontan undertaken; no complications reported during procedure.	PV oversewn at SCPC/Fontan	Readmission for decreased cardiac function & desaturation, placed on ECMO	Autopsy report: nonspecific findings, multiple pulmonary hemorrhages; Death after Fontan
1b	SPS (60)	0.25	Open	Death before SCPC	n/a	Genetic syndrome with multiple noncardiac abnormalities	Parents denied further palliation
1a*	SPS (83)	8.5	Closed	− Antegrade PBF/pulmonary atresia Bilateral BPA plasty at SCPC	n/a	Developed PLE after Fontan	Died from complications secondary to PLE and ultimate cardiac arrest; death after Fontan
1b	SPS (11)	1.6	Closed	− Antegrade PBF	MPA ligated at SCPC	—	Unspecified withdrawal of care
1b	SCPC (1.7 y)	2.2	Closed	− Antegrade PBF	PV oversewn at SCPC	Family returned to Haiti	Unspecified cause of death

(Continued)

TABLE E4. Continued

RVOT	Initial operation (age [d])	Age of death (y)	PDA status after first procedure	PBF status after SCPC	Details of MPA closure	Complications/morbidities	Details related to death
1b	SPS (5)	0.35	Open	Death before SCPC	n/a	Multiple congenital anomalies 2/2 to diabetic embriopathy Brought to ED unresponsive, identified stroke and seizure	Cardiac arrest during transport to hospital; "possible complications of dehydration"
1c	SPS (45)	1	Closed	Death before SCPC	MPA ligated at SPS	Readmission for acute respiratory distress	MSOF
1b	SPS (3)	0.6	Closed	− Antegrade PBF	MPA band (3 mm) at SPS; MPA ligated at SCPC	Return to OR after SCPC for takedown of SCPC and placement of BT shunt 2/2 cyanosis	Unspecified cause of death
1b	SPS (32)	0.13	Closed	Death before SCPC	n/a	Balloon atrial septectomy post- BT	Unspecified cause of death
1c	SPS (36)	0.24	Closed	Death before SCPC	n/a	—	Acute respiratory distress and cardiac failure
1c*	PAB (67)	1.15	Closed	− Antegrade PBF	MPA ligated at SCPC	—	Death after Fontan
1a	SPS (7)	0.12	Open, PGE continued	Death before SCPC/ pulmonary atresia	n/a	Readmission for acute desaturation POD 37; taken to OR for shunt revision (ductus ligated); + clot with neointimal hyperplasia found in shunt; Placed on inotropes-> ECMO	Withdrawal of care after shunt revision
1c*	SPS (2)	3.9	Closed	− Antegrade PBF	MPA ligated at SPS	Redo BT for thrombus in shunt	MSOF—death after Fontan
1c	PAB (68)	0.44	Closed	− Antegrade PBF	MPA ligated at SCPC	Thrombus in right atrium noticed on echocardiogram before death	Placed on ECMO after cardiac arrest
1b	SCPC (87)	5.4	Closed	+ Antegrade PBF	MPA banded at SCPC	—	Cardiac failure
1b*	SPS (5)	3.4	Closed	+ Antegrade PBF	PV oversewn at Fontan	Dysmorphic features, not thought to contribute to death	Death <30 d post-Fontan secondary to low cardiac output
1a*	SPS (6)	3.0	Closed	Pulmonary atresia	n/a	Developed PLE after Fontan	Died from cardiopulmonary arrest, after Fontan
1b	SPS (14)	<30 d	Closed	Death before SCPC	n/a	Developed NEC post-SPS	Withdrawal of care
1a	SPS (6)	<30 d	Closed	Death before SCPC/ pulmonary atresia	n/a	—	Unspecified cause of death in hospital after extubation

(Continued)

TABLE E4. Continued

RVOT	Initial operation (age [d])	Age of death (y)	PDA status after first procedure	PBF status after SCPC	Details of MPA closure	Complications/morbidities	Details related to death
1b	No surgery	—	—	—	—	—	—
1a	SPS (6)	1.3	Closed	Pulmonary atresia	n/a	—	Autopsy report: cardiomyopathy 2/2 to insufficient coronary artery perfusion
1b	SPS (4)	1.2	Closed	+ Antegrade PBF	MPA uninterrupted	—	Died at home; unspecified cause of death
1c	SCPC (132)	0.53	Closed	− Antegrade PBF	MPA ligated at SCPC	BT shunt after SCPC 2/2 to decreased O2 saturation	Unable to wean from ECMO
1c	SPS (95)	0.3	Closed	Death before SCPC	n/a	Placed on ECMO postoperatively	Death 1 mo after operation 2/2 to cardiac failure
1c	SPS (10)	1.2	Closed	− Antegrade PBF Bilateral BPA plasty at SCPC	MPA ligated at SPS	Readmitted for sepsis	MSOF and withdrawal of care
1b	SPS (11)	0.42	Closed	Death before SCPC	n/a	BT shunt stenosis with stent placed in shunt in cath lab	ECMO; withdrawal of care
1a	SPS (5)	0.84	Closed	Pulmonary atresia	n/a	—	Unspecified cause of death
1b	SPS (22)	1.6	Closed	− Antegrade PBF Bilateral BPA plasty at SCPC	PV oversewn at SPS	—	Unspecified cause of death
1c	PAB (44)	1.3	Closed	No antegrade PBF after SCPC	MPA ligated at SCPC	Reoperation after initial PAB for adjustment	Withdrawal of care; unspecified cause of death

Table shows details of PDA status, MPA closure, associated complications or morbidities, and cause of death, if available. If the status of postoperative PGE was noted in the operative note, it is listed here. (−) Antegrade PBF denotes no antegrade PBF throughout the MPA after SCPC; + antegrade PBF denotes any PBF after SCPC, throughout the MPA. *RVOT*, Right ventriular outflow tract; *PDA*, patent ductus arteriosus; *PBF*, pulmonary blood flow; *SCPC*, superior cavopulmonary anastomosis operation; *MPA*, main pulmonary artery; O_2, oxygen; *SPS*, systemic to pulmonary artery shunt operation; *n/a*, not applicable; *PGE*, prostaglandin infusion; *POD*, postoperative day; *BPA*, branch pulmonary artery; *PV*, pulmonary valve; *ECMO*, extracorporeal membrane oxygenation; *OR*, operating room; *PLE*, protein-losing enteropathy; *ED*, emergency department; *MSOF*, multisystem organ failure; *BT*, Blalock-Taussig; *PAB*, pulmonary artery band; *NEC*, necrotizing enterocolitis. *Death after Fontan palliation. If mentioned in the operative note, the type of MPA closure was classifed as closure at the level of the PV (PV oversewn) or MPA ligation, which refers to a nonspecific MPA closure. RVOT morphology based on institutional echocardiogram reports at diagnosis.

000 Surgical management of competing pulmonary blood flow affects survival before Fontan/Kreutzer completion in patients with tricuspid atresia type I

Travis J. Wilder, MD, Gerhard Ziemer, MD, PhD, Edward J. Hickey, MD, Peter J. Gruber, MD, PhD, Tara Karamlou, MD, MSc, Paul M. Kirshbom, MD, Eugene H. Blackstone, MD, William M. DeCampli, MD, PhD, William G. Williams, MD, and Brian W. McCrindle, MD, MPH, Toronto, Ontario, Canada; Chicago, Ill; Iowa City, Iowa; San Francisco, Calif; Hartford and New Haven, Conn; Cleveland, Ohio; and Orlando, Fla

For children with tricuspid atresia, judicious management of competing pulmonary blood flow may improve survival to Fontan operation.