Hybrid Versus Norwood Strategies for Single-Ventricle Palliation

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Background—Hybrid and Norwood strategies differ substantially in terms of stage II palliative procedures. We sought to compare these strategies with an emphasis on survival and reintervention after stage II and subsequent Fontan completion.

Methods and Results—Of 110 neonates with functionally single-ventricle physiology who underwent stage I palliation between 2004 and 2010, 75 (69%) infants (Norwood, n=43; hybrid, n=32) who subsequently underwent stage II palliation were studied. Survival and reintervention rates after stage II palliation, anatomic and physiologic variables at pre-Fontan assessment, and Fontan outcomes were compared between the groups. Predictors for reintervention were analyzed. Freedom from death/transplant after stage II palliation was equivalent between the groups (Norwood, 80.4% versus hybrid, 85.6% at 3 years, $P=0.66$). Hybrid patients had a higher pulmonary artery (PA) reintervention rate ($P=0.003$) and lower Nakata index at pre-Fontan evaluation ($P=0.015$). Aortic arch and atrioventricular valve reinterventions were not different between the groups. Ventricular end-diastolic pressure, mean PA pressure, and ventricular function were equivalent at pre-Fontan assessment. There were no deaths after Fontan completion in either group (Norwood, n=25, hybrid, n=14).

Conclusions—Survival after stage II palliation and subsequent Fontan completion is equivalent between the groups. The hybrid group had a higher PA reintervention rate and smaller PA size. Both strategies achieved adequate physiology for Fontan completion. Evolution of the hybrid strategy requires refinement to provide optimal PA growth. **(*Circulation*. 2012;126[suppl 1]:S123–S131.)**

Key Words: hypoplastic left heart syndrome ■ single ventricle ■ hybrid ■ Norwood ■ pulmonary artery

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The goal of staged surgical palliation for patients with functionally single-ventricle physiology is to produce the optimal Fontan candidate. The Norwood strategy has recently been evaluated in terms of the use of a Blalock-Taussig (BT) or a right ventricle–pulmonary artery (RV-PA) shunt.[1] At the time of stage II palliation, the Norwood procedure performed using either shunt type converges on a common stage II palliative procedure with a low incidence of morbidity and mortality.[1] The hybrid strategy has recently been introduced as an alternative surgical palliation strategy.[2–4] In contrast to the Norwood strategy, the stage II hybrid procedure is a "big operation," including aortic arch reconstruction, PA debanding and reconstruction, and creation of a bidirectional cavopulmonary shunt (BCPS).[3,4] After stage II procedures, both strategies converge on a common Fontan procedure and therefore, a comparison at pre-Fontan evaluation is germane.

We have reported equivalent hemodynamics and PA growth between Norwood and hybrid groups at pre–stage II palliation[5]; however, the impact of hybrid palliation on anatomic and physiologic variables after stage II palliation are less well defined. Potential concerns after stage II hybrid procedures include distortion of the branch PAs, aortic arch obstruction after removal of the ductal stent and subsequent arch reconstruction, and loss of ventricular function or increase in atrioventricular (AV) valve insufficiency due to the requisite long cardioplegic cardiac arrest during stage II palliation. We sought to compare our hybrid and Norwood experience with a specific focus on survival and reintervention after stage II palliation, anatomic and physiologic variables at pre-Fontan assessment, and subsequent Fontan outcome.

Methods

All infants undergoing staged single-ventricle palliation at the Hospital for Sick Children between 2004 and 2010 were reviewed. Research ethics board approval was obtained. During the study period, 119 consecutive infants underwent stage I single-ventricle palliation with hybrid (n=47), Norwood with a modified Blalock-Taussig (BT) shunt (n=63), or Sano procedure (n=9). The patients

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Table 1. Perioperative Data at Stage II Palliation

	Norwood (n=43)	Hybrid (n=32)	*P*
Patient demographics			
Age, mo	5.95 (4.81–6.93)	6.34 (5.72–7.24)	0.084
Body weight, kg	6.60 (5.90–7.22)	6.39 (5.54–7.38)	0.692
Body surface area, m^2	0.37 (0.34–0.40)	0.37 (0.34–0.41)	0.946
Diagnosis			
Hypoplastic left heart syndrome	23 (53%)	22 (69%)	0.182
Aortic atresia/mitral atresia	3 (7%)	11 (34%)	0.007
Aortic atresia/mitral stenosis	6 (14%)	3 (9%)	0.297
Aortic stenosis/mitral atresia	2 (5%)	0 (0%)	0.157
Aortic stenosis/mitral stenosis	12 (28%)	8 (25%)	0.286
Double-inlet left ventricle, transposition of the great arteries	8 (19%)	1 (3%)	0.041
Tricuspid atresia, transposition of the great arteries	4 (9%)	2 (6%)	0.063
Unbalanced atrioventricular septal defect	1 (2%)	4 (13%)	0.081
Double-outlet right ventricle, coarctation of the aorta	2 (5%)	2 (6%)	0.761
Other diagnoses	5 (12%)	1 (3%)	0.179
Secondary diagnosis: bilateral superior vena cava	1 (2%)	5 (16%)	0.036
Preoperative data			
Preoperative ECMO	2 (5%)	2 (6%)	0.761
Atrioventricular valve regurgitation			
None/mild	14 (33%)/15 (35%)	20 (63%)/5 (16%)	0.010/0.062
Moderate/severe	14 (33%)/0 (0%)	6 (19%)/1 (3%)	0.181/0.243
Ventricular function			
Normal/mildly reduced	34 (79%)/5 (12%)	28 (88%)/3 (9%)	0.340/0.755
Moderately reduced	4 (9%)	1 (3%)	0.289
Concomitant procedure			
Valve repair	10 (23%)	3 (9%)	0.116
Pulmonary vein stenosis repair	2 (5%)	1 (3%)	0.739
Intraoperative data			
Cardiopulmonary bypass time, min	85.0 (59.5–127.0)	207.5 (183.5–276.0)	<0.001
Aortic cross-clamp time, min	53.0 (23.0–138.0)	123.5 (95.8–152.3)	0.002
Circulatory arrest time, min	43.5 (11.3–64.5)	29.0 (20.0–45.0)	0.673
Regional cerebral perfusion, n	2 (5%)	31 (97%)	0.000
Regional cerebral perfusion time, min	28.0 (17.0–39.0)	58.0 (43.0–74.0)	0.211
Pulmonary artery plasty, n	37 (86%)	30 (94%)	0.285
Second pump run, n	3 (7%)	11 (34%)	0.003
Open chest, n	6 (14%)	9 (28%)	0.129
Postoperative data			
Central venous pressure, mm Hg	16.5 (15.0–18.0)	18.0 (15.5–19.5)	0.093
Minimum mixed venous saturation after stage II, %	40.0 (28.0–46.0)	33.0 (27.5–39.3)	0.031
Postoperative ECMO	2 (5%)	2 (6%)	0.761
Atrioventricular valve regurgitation			
None/mild	10 (23%)/18 (42%)	15 (47%)/9 (28%)	0.032/0.220
Moderate/severe	14 (33%)/1 (2%)	8 (25%)/0 (0%)	0.477/0.385
Ventricular function			
Normal/mildly reduced	29 (67%)/8 (19%)	19 (59%)/6 (19%)	0.472/0.987
Moderately reduced/severely reduced	4 (9%)/1 (2%)	5 (16%)/2 (6%)	0.405/0.391
Intubation time, d	1.0 (0.0–4.5)	2.5 (1.0–9.3)	0.054
Intensive care unit time, d	5.0 (2.0–13.5)	7.0 (3.0–17.3)	0.145
Hospital time, d	9.0 (7.0–19.0)	16.0 (9.8–28.3)	0.041

ECMO indicates extracorporeal membrane oxygenation.

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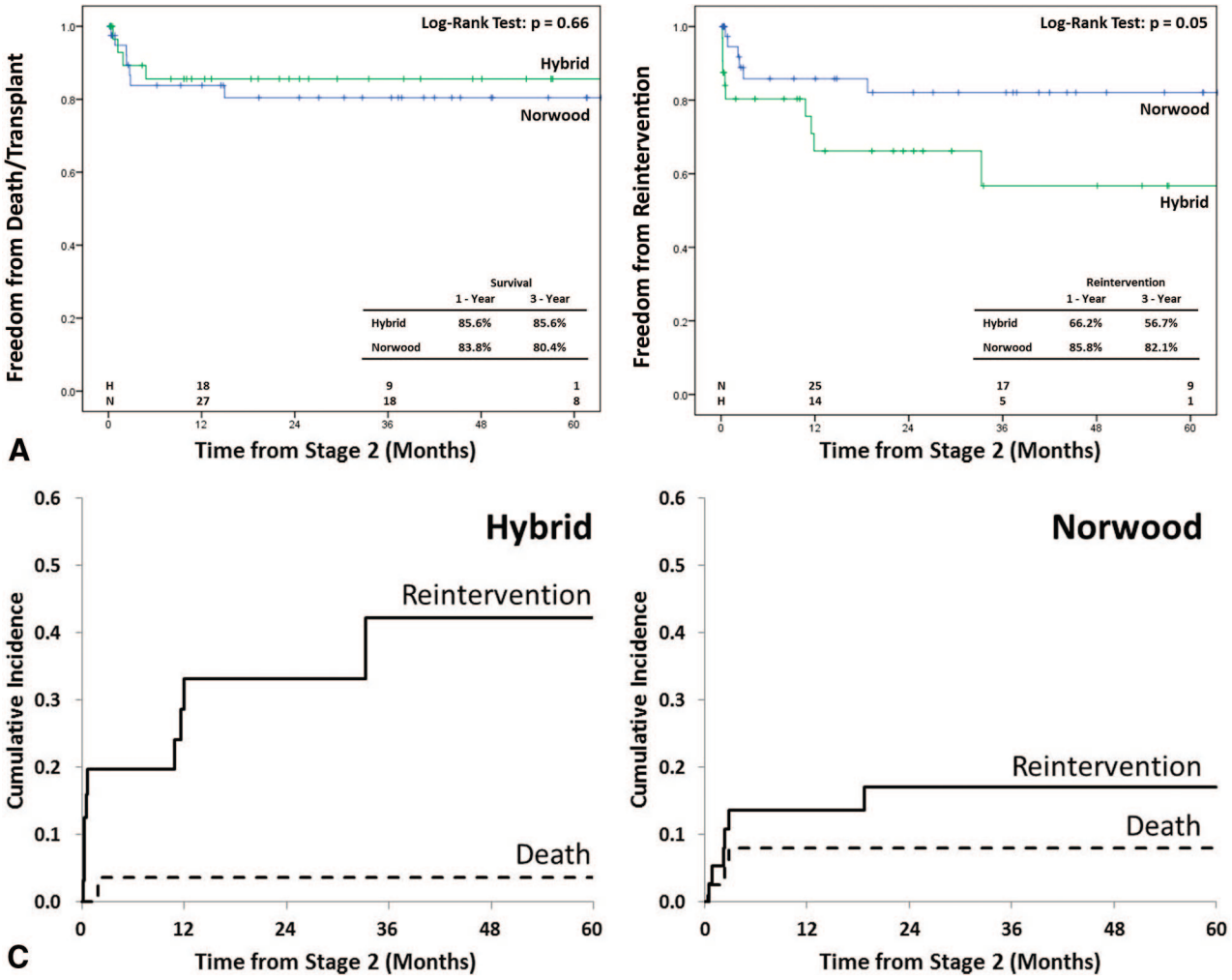

Figure 1. Kaplan-Meier analyses for freedom from death/transplant (**A**) and reintervention (**B**) after stage II palliation. Competing risk analysis showing outcomes after stage II palliation (**C**).

who underwent Sano procedure using RV-PA shunt were excluded from the analysis. This study analyzed 75 (68%) patients (43 Norwood, 32 hybrid) who subsequently underwent stage II palliation. Diagnoses and preoperative profile are shown in Table 1.

Surgical Techniques

The surgical techniques for stage I Norwood[6] or hybrid[4] palliative procedures have been previously described.

Norwood Stage II Procedure

After aorto-bicaval cannulation, the BT shunt was divided. Bilateral PA plasty was commonly performed with autologous pericardium. The superior vena cava (SVC) was anastomosed to the central portion of the right PA. If there was a left SVC, bilateral BCPS was performed as centrally as possible making a V-shaped appearance to minimize the central PA hypoplasia.[7]

Hybrid Stage II Procedure

Cardiopulmonary bypass was established with the arterial inflow through the main PA and ascending aorta (if large enough) and bicaval cannulation, the branch PAs occluded, and the patient cooled for deep hypothermic circulatory arrest (DHCA). After cardioplegic cardiac arrest, an atrial septectomy was performed. Under DHCA and regional cerebral perfusion, the ascending aorta was transected, the PA origins resected, the ductal stent was removed, and the aortic arch reconstructed with pulmonary homograft patch in a manner similar to Norwood stage I procedures. Alternatively, some patients had the aortic arch reconstruction using a retained stented duct (n=12).[8] In those cases, stent material was resected as the incision was made on the superior aspect of the arterial duct down to the posterior aspect of the proximal descending aorta. The remaining portion of stented duct at its lesser curvature was used as a part of aortic arch reconstruction. The proximal Damus-Kaye-Stansel (DKS) procedure was performed using the standard technique. Branch PAs were debanded and typically enlarged with fresh autologous pericardium from hilum to hilum. BCPS was completed while rewarming.

Since 2009, all comprehensive stage II palliations have been performed in the hybrid suite and exit angiograms have been routinely obtained at the end of operation. After termination of cardiopulmonary bypass (CPB), a 6F sheath is inserted via the SVC cannulation site. A 4F catheter is inserted into the lower SVC and an angiogram including BCPS anastomosis and branch PA architecture is obtained. The angiogram is immediately assessed by surgeons and cardiologists and decisions for reintervention are made.

Fontan Completion

The surgical techniques were essentially the same between the two groups. Extracardiac Fontan connections were created with 20- to 24-mm polytetrafluoroethylene tube grafts in the on-pump beating state with aortic and bicaval cannulation; 4-mm fenestrations were routinely created.

PAs and Aortic Arch

The angiographic images at pre-Fontan evaluation were reviewed. Diameters of branch PAs were measured at the hilum proximal to the

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Table 2. Reintervention After Stage II Palliation

	Norwood (n=43)	Hybrid (n=32)	P
Catheter intervention			
Pulmonary artery stent/dilatation	0 (0%)	6 (19%)	0.003
Coarctation dilation	0 (0%)	1 (3%)	0.243
Superior vena cava dilation	1 (2%)	2 (6%)	0.391
Surgical interventions			
Pulmonary artery angioplasty	1 (2%)	4 (13%)	0.081
Arch repair	1 (2%)	2 (6%)	0.391
Atrioventricular valve repair/replacement	2 (5%)	2 (6%)	0.761
Pulmonary vein stenosis repair	0 (0%)	1 (3%)	0.243
Additional Blalock-Taussig shunt	2 (5%)	0 (0%)	0.216
Bidirectional cavopulmonary shunt takedown	1 (2%)	2 (6%)	0.391
Transplantation	1 (2%)	1 (3%)	0.832

takeoff of the branching vessels. The Nakata index was calculated.[9] Diameters of lower lobe arteries were measured distal to their origin. The lower lobe index (LLI) was calculated.[5] The aortic diameters were measured: the neoascending aorta proximal to the innominate artery, the transverse aortic arch at the base of the left common carotid artery, the descending aorta at the level of the diaphragm, the ascending aorta distal to the sinotubular junction. Six patients (4 Norwood, 2 hybrid) underwent magnetic resonance imaging as a modality of anatomic assessment combined with catheter-based pressure measurement and were included in the analysis.

Assessment of Ventricular and AV Valve Function

Echocardiographic images at pre-Fontan evaluation were rereviewed. Qualitative assessments of ventricular function and degree of AV valve regurgitation were performed as previously described.[10] Ventricular function was graded as 0=normal, 1=mildly reduced, 2=moderately reduced, and 3=severely reduced. The degree of AV valve insufficiency was graded as 0=none/trivial, 1=mild, 2=moderate, and 3=severe.

Statistical Analysis

Continuous data are presented as median (interquartile range). Discrete data are presented as frequency (percentage). The level of statistical significance was set at $P \leq 0.05$. Differences between the groups were analyzed with the Mann-Whitney *U* test. Frequencies of the events were compared with the χ^2 test. Freedom from death/reintervention were analyzed with Kaplan-Meier analysis and a log-rank test. Cumulative incidence functions were used to display the proportion of patients with the event of interest (reintervention) or the competing event (death) as time progressed. Univariable predictors for reintervention were explored with Cox regression. Variables that were significant at the $P \leq 0.05$ level in univariable analysis were included in a stepwise multivariable Cox regression model.

Results

Stage II Palliation

Patient characteristics at stage II palliation were comparable between the groups. Hybrid stage II procedures were associated with longer CPB time ($P<0.001$) and the use of DHCA. Bilateral PA plasty was common in both groups (Norwood, 86% versus hybrid, 94%, $P=0.29$). Second bypass runs to address residual lesions were more common in the hybrid group (7% versus 34%, $P=0.003$). In the hybrid group, second bypass runs were required to revise branch PAs (n=3), BCPS anastomosis (n=3), DKS anastomosis (n=2), the aortic arch (n=1), and to achieve hemostasis (n=2). In the Norwood group, second bypass runs were required to revise AV valve repairs (n=3). There was a significant difference in lower minimum mixed venous saturations ($P=0.031$) as well as nonsignificant trends of higher SVC pressure ($P=0.093$) and longer intubation time ($P=0.054$) in the hybrid group. The length of hospital stay was longer in the hybrid group ($P=0.041$) (Table 1).

Survival

There were 4 (9%) hospital deaths in the Norwood group: cardiac arrest (n=2), hypoxia (n=1), and hemodynamic compromise after BCPS takedown (n=1). BCPS in all 4 patients had been undertaken after long intensive care unit stays after the Norwood procedure. There were 2 (6%) hospital deaths in the hybrid group: multi-organ failure after BCPS takedown (n=1), and PA distortion and subsequent extracorporeal membrane oxygenation (ECMO) support, balloon angioplasty, BCPS takedown, and multi-organ failure (n=1). There were 3 late deaths in the Norwood group: cardiac arrest (n=1), respiratory failure (n=1), and sudden death (n=1). There was 1 late death in the hybrid group due to pneumonia. Two patients (1 patient in each group) underwent cardiac transplantation after stage II palliation. Both patients had reduced ventricular function prior to stage II palliation.

Freedom from death/transplant after stage II palliation was comparable between groups at 1 year (Norwood, 83.8% versus hybrid, 85.6%) and at 3 years (80.4% versus 85.6%) (log-rank test, $P=0.66$) (Figure 1A).

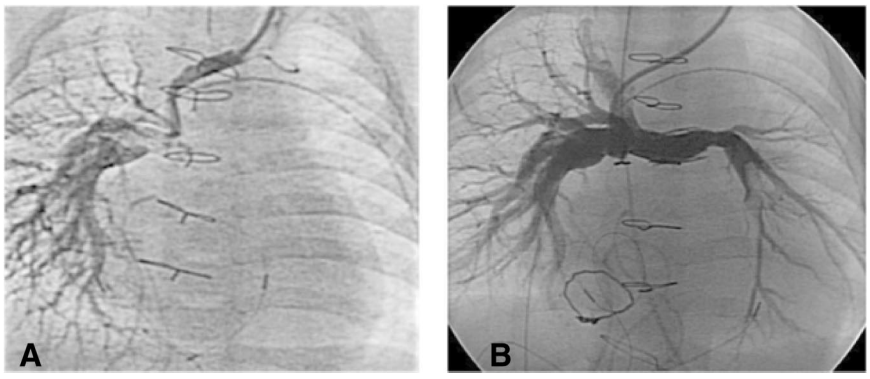

Figure 2. Occlusion of left pulmonary artery after comprehensive stage II hybrid palliation (**A**). This was treated with stent placement (**B**).

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Table 3. Pre-Fontan Evaluation

	Norwood (n=25)	Hybrid (n=14)	*P*
Patient demographics			
Body weight, kg	12.7 (11.9–13.2)	11.6 (11.2–13.2)	0.195
Body surface area, m^2	0.55 (0.52–0.57)	0.53 (0.51–0.55)	0.185
Catheterization/magnetic resonance imaging data			
Arterial saturation, %	88.0 (84.8–90.3)	89.0 (87.0–91.0)	0.236
Ventricular end-diastolic pressure, mm Hg	7.0 (6.0–8.0)	7.5 (7.0–8.0)	0.665
Mean pulmonary artery pressure, mm Hg	10.0 (9.0–12.0)	10.0 (9.0–11.5)	0.605
Common atrial pressure, mm Hg	5.5 (4.3–6.8)	5.0 (4.5–6.0)	0.611
Transpulmonary gradient, mm Hg	5.0 (4.0–6.0)	5.3 (4.3–6.0)	0.981
Pulmonary-to-systemic flow ratio, Qp/Qs	0.81 (0.74–1.02)	1.10 (0.88–1.19)	0.383
Pulmonary vascular resistance, Woods Units	2.36 (1.94–2.70)	2.20 (1.95–2.95)	0.877
Pulmonary arteries			
Right pulmonary artery diameter, mm	9.90 (9.05–11.53)	8.35 (6.80–9.50)	0.003
Indexed right pulmonary artery diameter, mm^2/m^2	18.5 (17.0–20.5)	15.0 (13.5–16.7)	0.008
Left pulmonary artery diameter, mm	7.40 (6.45–8.20)	6.20 (5.43–7.50)	0.071
Indexed left pulmonary artery diameter, mm^2/m^2	13.3 (11.7–15.2)	13.1 (10.7–13.9)	0.273
Nakata Index, mm^2/m^2	216 (187–277)	184 (127–213)	0.015
Right-to-left ratio	1.43 (1.24–1.63)	1.15 (1.10–1.65)	0.622
Lower lobes			
Right lower lobe diameter, mm	7.45 (5.98–8.23)	6.10 (5.68–6.85)	0.093
Left lower lobe diameter, mm	6.20 (5.23–6.60)	5.25 (4.20–6.10)	0.087
Lower lobe index, mm^2/m^2	134 (96–156)	109 (75–131)	0.113
Aorta			
Neo-ascending aorta, mm	22.6 (18.6–25.0)	18.7 (17.1–21.0)	0.035
Distal transverse arch, mm	15.5 (13.9–18.6)	16.1 (13.5–18.3)	0.936
Descending aorta, mm	8.60 (8.05–9.35)	7.90 (7.33–9.23)	0.208
Original ascending aorta, mm	8.10 (5.70–9.90)	7.05 (4.45–8.48)	0.360
PA window, mm	20.4 (17.8–25.0)	18.7 (17.3–22.5)	0.257
Echocardiographic data			
Atrioventricular valve regurgitation			
None	10 (40%)	8 (57%)	0.303
Mild	11 (44%)	5 (36%)	0.614
Moderate	4 (16%)	1 (7%)	0.427
Ventricular function			
Normal	24 (96%)	13 (93%)	0.669
Mildly reduced	1 (4%)	1 (7%)	0.669

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Reintervention

After stage II palliation, 20 reinterventions were performed in 10 (31%) patients in the hybrid group and 8 reinterventions in 5 (11%) patients in the Norwood group, at median of 0.56 months (interquartile range, 0.36–11.17 months) after surgery (Table 2). Balloon angioplasty with/without stent placement for the PA was performed in 6 (19%) patients in the hybrid group and none in the Norwood group ($P=0.003$) (Figure 2). Reintervention rates for the aortic arch and AV valves were similar between the groups. Freedom from all reinterventions was higher in the Norwood group at 1 year (85.8% versus 66.2%) and at 3 years (82.1% versus 56.7%) (log-rank test, $P=0.05$) (Figure 1B and 1C). Cox multivariable regression showed longer cardiopulmonary bypass time ($P=0.012$), longer intubation time ($P=0.009$), longer hospital time ($P=0.016$), and reduced ventricular function after stage 2 ($P=0.016$) as predictors for reintervention.

Pre-Fontan Evaluation

Arterial saturation, ventricular end-diastolic pressure, mean PA pressure, pulmonary-to-systemic flow ratio (Qp/Qs), and pulmonary vascular resistance were comparable between the groups (Table 3). The diameter of the bilateral PAs and Nakata index are larger in the Norwood group ($P=0.015$) (Figure 3 and Table 3). There were no significant differences in aortic dimensions except for a smaller neoascending aorta in the hybrid group

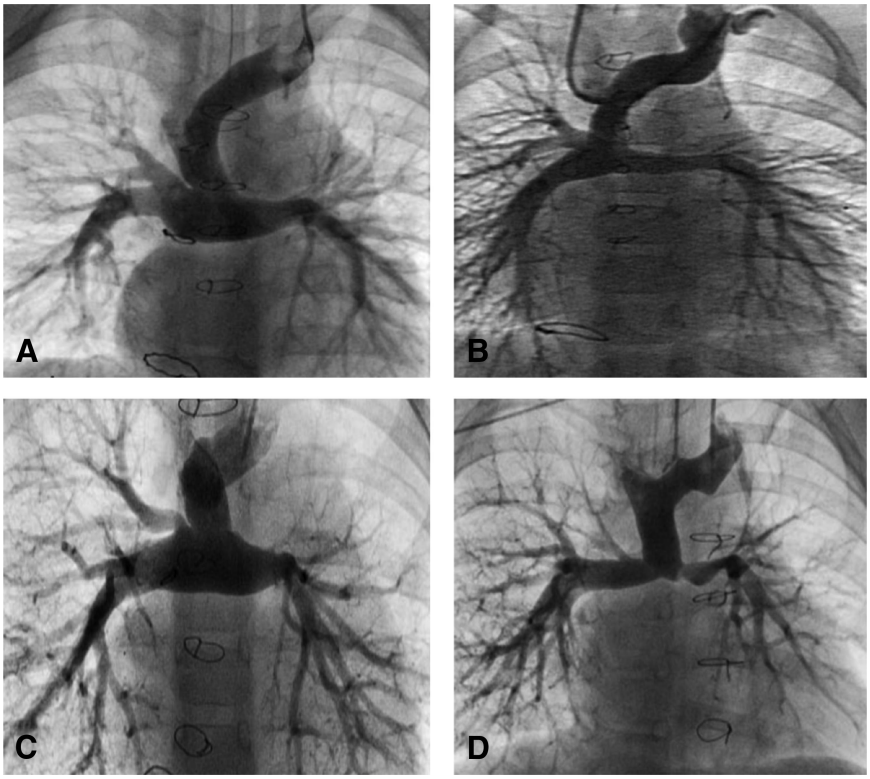

Figure 3. Well-developed symmetrical branch pulmonary arteries (PAs) (**A**) and hypoplastic left PA (**B**) in a Norwood patient at pre-Fontan assessment. Well-developed symmetrical branch PAs (**C**) and hypoplastic left PA (**D**) in a hybrid patient.

($P=0.035$) (Figure 4). Most patients in both groups had normal echocardiographic ventricular function (Norwood, 96% versus hybrid, 93%, $P=0.67$) and mild or less AV valve regurgitation (84% versus 93%, $P=0.43$). Four patients in the Norwood group and 1 patient in the hybrid group had moderate AV valve regurgitation, and there were no patients who had severe regurgitation in either group.

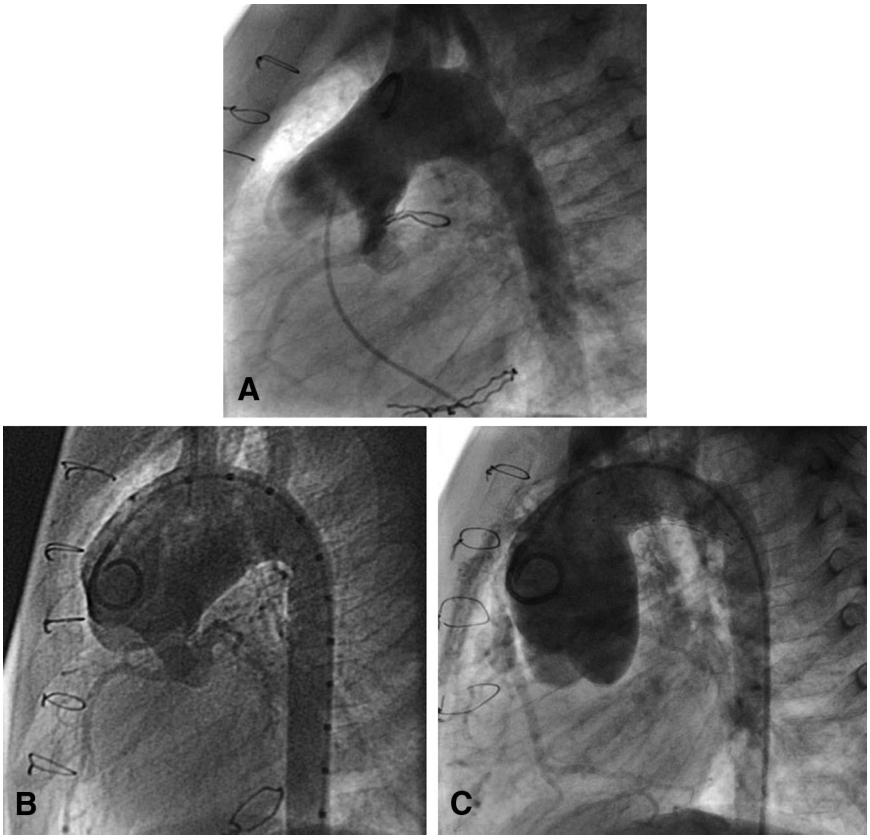

Figure 4. Aortogram of: a Norwood patient (**A**); standard arch reconstruction in a hybrid patient (**B**); arch reconstruction in a hybrid patient with a retained stented duct (**C**).

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Table 4. Perioperative Data at Fontan Completion

	Norwood (n=25)	Hybrid (n=14)	*P*
Patient demographics			
Age, mo	37.0 (31.8–41.2)	36.3 (32.1–41.9)	0.761
Time after stage II palliation, mo	29.8 (25.6–32.7)	30.7 (23.6–34.8)	0.675
Body weight, kg	13.7 (13.0–14.7)	13.7 (12.7–14.6)	0.927
Body surface area, m^2	0.55 (0.53–0.57)	0.55 (0.53–0.57)	0.832
Concomitant procedure			
Pulmonary artery plasty	2 (8%)	6 (43%)	0.010
Valve repair	2 (8%)	0 (0%)	0.277
Intraoperative data			
Cardiopulmonary bypass time, min	40.0 (34.0–61.0)	83.5 (46.5–101.3)	0.013
Aortic cross-clamp, n	6 (24%)	0 (0%)	0.046
Central venous pressure, mm Hg	15.0 (13.5–17.0)	14.0 (12.5–16.0)	0.300
Postoperative data			
Intubation time, d	0.0 (0.0–1.0)	0.0 (0.0–1.0)	0.976
Intensive care unit time, d	2.0 (1.0–4.0)	2.0 (1.0–3.0)	0.649
Hospital time, d	9.0 (7.0–17.0)	14.0 (11.0–34.0)	0.074
Postoperative morbidity/reintervention			
Chylothorax	4 (16%)	7 (54%)	0.015
Coil occlusion/embolization of collaterals	3 (12%)	5 (38%)	0.058
Thoracic duct ligation	0 (0%)	2 (15%)	0.111
Balloon angioplasty and stent	0 (0%)	1 (8%)	0.160
Protein-losing enteropathy	1 (4%)	1 (8%)	0.629
Pacemaker	0 (0%)	1 (8%)	0.160
Superior vena cava/inferior vena cava clot	1 (4%)	0 (0%)	0.465

Fontan Outcome

Age and body size at Fontan completion were comparable between the groups (Table 4). The hybrid group had a longer CPB time ($P=0.013$) and higher PA plasty rate (Norwood, 8% versus hybrid, 43%, $P=0.010$). There were no differences in central venous pressure, intubation time and intensive care unit stay between the groups. There was a nonsignificant trend toward longer length of hospital stay in the hybrid group ($P=0.074$), which may be related to a higher chylothorax rate ($P=0.015$).

Overall Outcome

The overall outcome of the entire cohort is shown in Figure 5. The 1- and 3-year survival of the entire cohort from birth was analyzed. Freedom from death or transplant was 63.7% at 1 year and 58.9% at 3 years for the Norwood group and 68.3% at 1 year and 65.1% at 3 years for the hybrid group (log-rank test, $P=0.71$). All patients in both groups who are awaiting Fontan completion are considered to be Fontan candidates. None of the patients were deemed to be unsuitable for Fontan completion at their latest follow-up.

Discussion

Hybrid palliation for neonates with single-ventricle physiology has evolved as a new surgical paradigm avoiding CPB and DHCA.[2–4] Because the key anatomic elements remain untouched at this stage, the comprehensive stage II hybrid palliation is more technically demanding, requiring reconstruction of both aortic arch and PA structures. The primary focus of this study was to define the impact of Norwood versus hybrid strategies on anatomic and physiologic parameters looking toward Fontan candidacy and subsequent clinical outcomes. The key findings include equivalent survival between the groups up to 3 years after stage II palliation, and higher PA reintervention and less developed PA structures in the hybrid group. These results underscore the significant challenges in reconstructing the branch PAs at stage II hybrid palliation. Ventricular and AV valve functions and overall pre- and post-Fontan physiology were comparable between the groups.

Survival

Survival after stage II palliation was comparable between the strategies. Most patients in the hybrid group tolerated the lengthy procedure involving DHCA and had equivalent mortality and postoperative recovery compared with the Norwood group, confirming the physiological tolerance of this age group. The hospital mortality (6%) of the comprehensive stage II palliation was similar to another series by Galantowicz et al.[11] Mortalities in this series partly reflect the learning curve of the new surgical strategy but were also related to residual PA stenosis. After experiencing the second mortality, we used a routine exit angiogram to visualize the PA and BCPS architecture to detect major residual lesions.

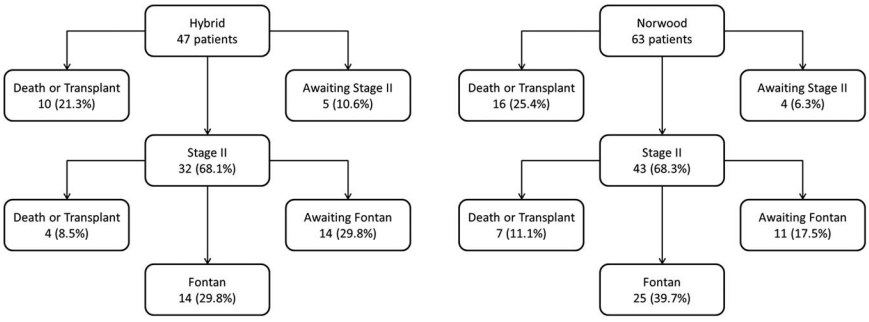

Figure 5. Overall outcomes for the hybrid and Norwood groups.

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The aortic arch reconstruction was not associated with mortality. The results emphasized that the quality of the PA reconstruction at stage II palliation was strongly related to morbidity and mortality.

Pulmonary Arteries

The most important finding was the significant challenge in reconstructing the PAs at comprehensive stage II palliation and in achieving subsequent reasonable PA architecture for Fontan completion. Many patients required revision of or reinterventions for the branch PAs, resulting in a lower Nakata index. Nonetheless, PA physiology was maintained with reasonable PA pressures and resistance, revealing no impact on Fontan outcome. The mechanism of branch PA stenosis mainly stems from anatomical distortion of the branch PAs, but also from the difficulty in geometric arrangement between the newly reconstructed aorta and branch PAs.

The staged single-ventricle palliations had encountered problems with the branch PAs. Left PA stenosis after stage I Norwood procedure was common in the past,[12] and was largely overcome by technical modifications.[12,13] Central branch PA stenosis has been common after stage I Sano procedures, and was partly but not entirely overcome by technical modifications.[14–15] Unlike those problems, the PA problem in hybrid palliation is unique because the problems occur at stage II palliation where the PAs are fed by a passive BCPS rather than an active BT shunt. The branch PAs can be easily compressed by the reconstructed aorta because of a low pressure profile. This fact leaves limited options other than an extensive PA patch plasty with aortic arch reconstruction wherein the aortic arch is reduced in size to minimize PA compression. Asymmetrical PA bands at the time of stage I palliation may have a negative impact on PA geometry and subsequent PA reconstruction. This problem has yet to be overcome and surgical technical modifications are of prime importance in improving PA outcomes.

Aortic Arch Reconstruction

The stented duct did not add any clinical risks or affect the quality of the aortic arch reconstruction in the hybrid group. The aortic arch reconstruction with stent removal or with the use of a retained stented duct was durable, and there was no mortality associated with the aortic arch reconstruction in the hybrid group. The reintervention rate on the reconstructed aorta (n=3, 9%) was not significantly higher than that in Norwood stage I palliation.[6,16] Galantowicz et al[11] reported no reintervention for the aortic arch in 36 patients after stage II palliation. The aortic architecture at pre-Fontan assessment was comparable between the groups. Of note, aortic arch reconstruction with a retained stented duct seemed to achieve equivalent aortic arch architecture without adding any adverse effects. Since the follow-up period is short, the aortic arch status in the hybrid group should be carefully monitored.

Ventricular and AV Valve Function

Our study clearly showed that both strategies equally preserved ventricular and AV valve function after stage II palliation. This is not surprising given that conventional and comprehensive stage II palliations result in the same physiology of "in-series" circulation with a volume-unloaded ventricle. Our previous study also showed that Norwood and hybrid stage I palliation resulted in similar volume overload, that is, Qp/Qs, at pre–stage II evaluation.[5] The only difference between the strategies in terms of the impact on ventricular function is the timing of the cardioplegic cardiac arrest. The result showed that there was no incidence of severe ventricular dysfunction after comprehensive stage II palliation. Our previous report showed somewhat higher rates of ventricular dysfunction and transplantation in the Norwood group after stage I palliation.[5] This may be partly explained by the fact that chronic cyanosis may increase tolerance to ischemia-reperfusion injury in developing hearts.[17] This may be an additional rationale to move towards hybrid palliation to avoid an ischemic insult on neonatal myocardium.

Fontan Candidacy and Clinical Outcome

During the process of conventional staged surgical palliation for patients with hypoplastic left heart syndrome, Fontan candidacy could be compromised by deterioration of ventricular function, AV valve function, or distortion of the branch PA architecture.[18–19] Theoretically, the hybrid palliation has similar risks. This study showed most Fontan candidates in the hybrid group had good ventricular and AV valve functions, and reasonable PA anatomy and physiology, making them reasonable candidates for Fontan completion. Death or transplant rate due to poor ventricular function was equivalent between the groups. Fontan completion was achieved with no mortality, although many patients (43%) required reaugmentation of the branch PAs, resulting in significantly longer CPB time. Postoperative parameters were equivalent between the groups except for a higher incidence of chylothorax in the hybrid group. Impact of stented branch PAs in the hybrid group on cavopulmonary physiology late after Fontan completion has yet to be determined.

Study Limitations

A major limitation of this study was its retrospective and nonrandomized nature, which could have resulted in a bias in patient selection between groups. Because the patients who underwent the Sano procedure as stage I palliation were excluded, the outcome of the Norwood group may have been altered. The follow-up period of this cohort is short (median follow-up, 22.0 months), and a limited proportion of both groups (Norwood, 58%; hybrid, 44%) have reached pre-Fontan assessment and subsequent Fontan operation. Data regarding pre-Fontan assessment and Fontan outcome may not represent the overall picture of this entity and therefore further follow-up studies are necessary.

Conclusions

Norwood and hybrid surgical strategies had equivalent survival after stage II palliation. The hybrid group had a significantly higher PA reintervention rate and subsequently, worse PA growth at pre-Fontan assessment. Despite having a small branch PA size, PA physiology in the hybrid group was adequate, having no impact on Fontan outcome. Surgical modifications in PA reconstruction are of prime importance in achieving further improvement in clinical outcomes.

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Disclosures

None.

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