Size of the right ventricle-to-pulmonary artery conduit impacts mid-term outcome after the Norwood procedure in patients weighing less than 3 kg

Naruhito Watanabe, MD, Petros V. Anagnostopoulos, MD, Takeshi Shinkawa, MD, Natalie Johnson, BS, and Anthony Azakie, MD

Objective: The optimal shunt size for patients who have the Norwood operation with a right ventricle-to-pulmonary artery conduit is controversial. The goal of this study is to compare outcomes of 2 shunt sizes in this population.

Methods: Between 2002 and 2010, 75 consecutive patients diagnosed with hypoplastic left heart syndrome and its variants underwent the Norwood procedure with a right ventricle-to-pulmonary artery conduit. The outcomes of 20 neonates weighing <3 kg were analyzed.

Results: The cumulative 30-day stage 1 mortality was 97% (95% confidence interval, 88%-99%) for all patients (73/75), 20 of whom weighed <3 kg. Nine patients had a 6-mm (group 1) and 11 patients had a 5-mm (group 2) right ventricle-to-pulmonary artery conduit. Thirty-day stage 1 survival was 88% (8/9) in group 1 and 90% (10/11) in group 2 ($P = .88$). The central pulmonary artery confluence size at prebidirectional cavo-pulmonary shunt catheterization was 4.5 ± 1.2 mm in group 1 and 2.5 ± 1.0 mm in group 2 ($P = .009$). The mean transpulmonary gradient was higher in group 2 (7.8 ± 3.1 mm Hg vs 4.2 ± 1.9 mm Hg; $P = .036$). The incidence of pulmonary artery intervention was 16% (1/7) in group 1 and 75% (6/8) in group 2 ($P = .030$). Survival rate at 6 months was 66% (6/9) in group 1 and 70% (7/10) in group 2 ($P = .89$).

Conclusions: Use of a 6-mm right ventricle-to-pulmonary artery conduit showed better central pulmonary artery growth and less need for pulmonary artery intervention in the authors' experience. (J Thorac Cardiovasc Surg 2012;144:1091-4)

The most successful palliation for hypoplastic left heart syndrome (HLHS) has been the Norwood procedure, first introduced in 1983.¹ The Norwood procedure has undergone several modifications and the outcomes of stage 1 palliation have improved significantly during the past few decades. Since Sano and colleagues² reported the use of the right ventricle-pulmonary artery (RV-PA) conduit, there has been much debate about its benefits and potential disadvantages. The main advantage of the RV-PA conduit is decreased diastolic runoff into the pulmonary vasculature. The higher systemic diastolic pressure compared with the modified Blalock-Taussig shunt (mBTS) results in improved myocardial perfusion with lower volume loading.³⁻⁷ The physiology with the RV-PA conduit results in more stable, immediate postoperative hemodynamics⁸⁻¹¹ A recent report of more than 500 infants undergoing the

Norwood procedure showed that after randomization, transplantation-free survival at 12 months was higher with the RV-PA conduit than with the mBTS.¹² However, some reported disadvantages of the RV-PA conduit include increased incidence of ventricular dysfunction, shunt stenosis, and central pulmonary artery (PA) stenosis.¹³⁻¹⁷ Moreover, precise evaluation of the affect of different RV-PA conduit sizes has not been done. We hypothesized that a 6-mm RV-PA conduit provided better PA growth, resolution of the problem, and satisfactory mid-term outcome compared with a 5-mm conduit. The purpose of this study was to identify the differences in surgical results, PA size, and the incidence of PA intervention after the stage 1 Norwood procedure with a 6-mm versus a 5-mm RV-PA conduit to help guide selection of shunt sizes for neonates with HLHS.

METHODS

Study Design

We performed a retrospective case-control study including all patients who had the Norwood procedure using an RV-PA conduit between April 2002 and December 2010 at our institution. The study was approved and monitored by the institutional review board and, because of its retrospective nature, the need for patient consent was waived. We analyzed the outcomes of stage 1 and stage 2 palliation in 20 neonates weighing <3 kg with HLHS or one of its variants. Patients were divided into either the 6-mm shunt group (group 1) or the 5-mm shunt group (group 2) on the basis of which size of an RV-PA conduit was placed at the time of the stage 1 reconstruction. The

From the Division of Pediatric Cardiothoracic Surgery, University of California San Francisco, San Francisco, Calif.

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Address for reprints: Anthony Azakie, MD, 513 Parnassus Ave, Med Sci S549, San Francisco, CA 94143-0117 (E-mail: Tony.Azakie@ucsfmedctr.org).
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Abbreviations and Acronyms

BCPS	=	bidirectional cavopulmonary shunt
HLHS	=	hypoplastic left heart syndrome
mBTS	=	modified Blalock–Taussig shunt
PA	=	pulmonary artery
RV-PA	=	right ventricle–pulmonary artery
SVC	=	superior vena cava

choice of shunt was at the discretion of the surgeon. The transition from the 5-mm RV-PA conduit to the 6-mm was in 2008. The primary study population comprised the total number of patients with pre-Glenn cardiac catheterization data available for analysis at our institution. Preoperative demographics and surgical data were compared for our study population. Surgical morbidity and mortality data through completion of the Glenn procedure were collected at follow-up.

Medical records were reviewed and the following data were retrieved and analyzed: basic demographic data; clinical presentation and past medical and surgical history; perioperative data, including associated surgical procedures; postoperative outcomes, including stage 1 Norwood procedure and stage 2 bidirectional cavopulmonary shunt (BCPS); echocardiographic data before stage 1 palliation, at hospital discharge, and before stage 2 palliation; and catheterization data before stage 2 palliation.

Surgical Procedure

Norwood procedure with RV-PA conduit. The stage 1 reconstruction for each patient was performed by the same surgeon. The standard Norwood procedure performed at our institution during the study period consisted of division of the main PA with side-to-side anastomosis to the ascending aorta, or homograft patch augmentation of the ascending aorta and aortic arch, with an RV-PA conduit, with atrial septectomy under selective cerebral perfusion. The final step involved the placement of a polytetrafluoroethylene conduit from the left side of the right ventricle infundibulum just below the pulmonary valve to the central PA. The RV-PA conduit was typically placed to the left of the neo-aorta and anastomosed directly to the glutaraldehyde-fixed pericardium patch on the PA bifurcation. No valved or ringed polytetrafluoroethylene grafts were used.

Off-pump BCPS. After repeat sternotomy, a test occlusion of the right PA was then performed. If hemoglobin–oxygen saturations remained $\geq 70\%$, then the off-pump BCPS¹⁸ was performed. The superior vena cava (SVC)–innominate vein junction and right atrial appendage were cannulated with 12-Fr Pacifico cannulas (DLP Inc, Grand Rapids, Mich) and were joined for insertion of a passive venous shunt. The SVC–right atrial junction was clamped and divided, and the cardiac end was oversewn with polypropylene sutures. The end of the SVC was sewn to the PA with running fine polypropylene sutures after proximal and distal control of the right PA was achieved. The azygous vein was ligated. The RV-PA conduit was then clipped and divided.

Catheterization Analysis

Pre-Glenn cardiac catheterizations performed at our institution were reviewed. The diameters of the right and left PAs were measured just proximal to the origin of the first-order PA branches and were used to calculate a Nakata index (combined right PA and left PA cross-sectional area indexed to body surface area). Shunt stenosis was defined as $>50\%$ narrowing of the shunt diameter. All measurements were corrected on the Vericis system (Camtronics Medical Systems, Hartland, Wis) and calibrated to known catheter or shunt diameter. The transpulmonary gradient was mean PA pressure less the mean left atrium pressure. Coronary perfusion pressure was the pressure gradient between the right atrial and aortic diastolic pressures.

Statistical Analysis

Demographic data, operative factors, and postoperative events were recorded. Continuous variables are presented as mean ($\pm$ standard deviation) for normally distributed data. For subgroup analyses, the Student *t* test was used to compare the parametric values; the Mann-Whitney *U* test was used to compare the nonparametric values. The 30-day survival, interstage mortality, and PA intervention rate were computed using the χ^2 test as categorical data. A probability value $<.05$ was considered significant for all analyses. Analysis was performed with Stat View 5.0 software (SAS Institute Inc, Cary, NC).

RESULTS

Patient Characteristics

There were no significant differences between the 2 groups for demographic data at the time of stage 1 Norwood procedure (Table 1). Mean body weight at stage 1 Norwood procedure was similar in both groups (2.6 ± 0.3 kg vs 2.5 ± 0.2 kg, $P = .46$). The mean age at stage 1 palliation was 7.5 ± 2.6 days in group 1 and 6.5 ± 3.4 days in group 2 ($P = .48$). No difference was observed for the presence of aortic atresia with mitral stenosis (11%, 1/9 vs 27%, 3/11; $P = .36$). The incidence of restrictive atrial septal defect was 22% (2/9) in group 1 and 18% (2/11) in group 2 ($P = .82$). No patient in either group had more than moderate tricuspid regurgitation. Group 2 included 1 patient with Noonan syndrome and 1 patient with trisomy 21.

Early and Interstage Results

Operative data are summarized in Table 1. The cumulative, 30-day, stage 1 mortality was 97% for all patients (73/75). Thirty-day stage 1 survival in patients weighing <3 kg was 88% (8/9) in group 1 and 90% (10/11) in group 2 ($P = .88$). Pulse oximetry oxygen saturation at discharge was $91.8 \pm 3.2\%$ in group 1 and $82.3 \pm 4.7\%$ in group 2 ($P = .0009$). No patient in either group developed severe ventricular dysfunction or arrhythmias at discharge. One patient had severe ventricular dysfunction at interstage in group 2. All patients were placed on an aspirin regimen postoperatively. There was no difference in the interstage mortality between the 2 groups (12%, 1/8 vs 10%, 1/10; $P = .49$), and 1 group 2 patient with aortic atresia with mitral stenosis and left ventricular sinusoids was referred for cardiac transplantation. Angiotensin-converting enzyme inhibitors were prescribed for 85% (6/7) in group 1 patients and 62% (5/8) in group 2 patients after stage 1 palliation ($P = .589$).

Mid-Term Results

The median age at BCPS was older in group 1 patients (194 days; range, 104–693 days vs 111 days; range, 72–172 days; $P = .028$), but weight was similar in both groups (5.2 ± 1.7 kg vs 4.9 ± 0.3 kg, $P = .78$; Table 2). Pulmonary/systemic flow ratio was 1.77 ± 0.88 in group 1 and 1.06 ± 0.39 in group 2 ($P = .05$). Ventricular end diastolic pressure was similar in both groups (9 ± 2 mm Hg vs 10 ± 4 mm Hg, $P = .60$). Mean

TABLE 1. Demographic and perioperative data at stage 1 palliation

	6-mm RV-PA conduit (n = 9)	5-mm RV-PA conduit (n = 11)	P value
Demographic data			
Age at stage 1, d	7.5 ± 2.6	6.5 ± 3.4	.48
Weight at stage 1, kg	2.6 ± 0.3	2.5 ± 0.2	.46
Male/female	3/6	7/4	.11
AA with MS	11% (1/9)	27% (3/11)	.36
Restrictive ASD	22% (2/9)	18% (2/11)	.82
Genetic syndrome	0% (0/9)	18% (2/11)	.17
Preoperative data at stage 1 Norwood			
Bypass time, min	113 ± 27.4	116.9 ± 33.8	.78
Crossclamp time, mn	41.2 ± 12.8	53 ± 11.5	.052
SpO ₂ at discharge, %	91.8 ± 3.2	82.3 ± 4.7	.0009

RV-PA, Right ventricle–pulmonary artery; AA, aortic atresia; MS, mitral stenosis; ASD, atrial septal defect; SpO₂, hemoglobin–oxygen saturation by pulse oximetry.

coronary perfusion pressure was higher in group 1 patients (39 ± 2 mm Hg vs 33 ± 6 mm Hg, $P = .026$). The mean central PA confluence size at pre-BCPS catheterization was 4.5 ± 1.2 mm in group 1 patients and 2.5 ± 1.0 mm in group 2 patients ($P = .009$). The diameter of the right PA was 8.7 ± 2.7 mm in the 6-mm shunt group and 6.8 ± 2.2 mm in the 5-mm shunt group ($P = .19$). The diameter of the left PA was 8.1 ± 3.0 mm in the 6-mm shunt group and 6.2 ± 3.1 mm in the 5-mm shunt group ($P = .30$). PA index was 416.6 ± 175.4 mm²/m² in group 1 and 349.2 ± 276.5 mm²/m² in group 2 ($P = .63$). The mean transpulmonary gradient was higher in group 2 patients (7.8 ± 3.1 vs 4.2 ± 1.9 mm Hg, $P = .036$). A balloon arterioplasty to the left PA in group 1

and a PA stent insertion to distal shunt stenosis in group 2 were performed. A left PA plasty and 5 bilateral PA plasty in group 2 were done at stage 2 palliation. The incidence of PA intervention (surgical or balloon arterioplasty ± PA stent insertion) was 16% (1/7) in group 1 and 75% (6/8) in group 2 ($P = .030$). All group 1 patients had BCPS without cardiopulmonary bypass, whereas no group 2 patient could have an off-pump BCPS ($P < .001$). No patient required tricuspid valve repair in either group. The 30-day survival after BCPS was 100% (6/6) in group 1 and 87% (7/8) in group 2 ($P = .36$). Survival rate at 6 months was 66% (6/9) in group 1 patients and 70% (7/10) in group 2 patients ($P = .89$). Neither group had RV-PA conduit obstruction at the proximal insertion site of the right ventricular infundibulum. Obstruction of the RV-PA conduit at the distal insertion site of the conduit into the PA bifurcation was identified in 1 group 1 patient and 2 group 2 patients.

DISCUSSION

Modifications to surgical technique and perioperative care have improved mortality and morbidity for patients with HLHS and its variants. The RV-PA modification to the Norwood procedure has become widely accepted as an effective alternative to the Norwood procedure with mBTS and represents an important development in surgical technique. The impact of the RV-PA conduit on PA size has been reported. Rumball and associates¹⁶ have shown that infants having the RV-PA modification of the Norwood procedure have larger left PA dimensions compared with those with the mBTS, but also have significant central PA hypoplasia. Pruetz and colleagues¹⁹ have also shown that the RV-PA conduit does result in significantly larger distal left PA size and more evenly balanced branch PA growth. However, there was a higher incidence of central PA hypoplasia and the need for additional shunting or early Glenn surgery. In a recent report by Griselli and co-workers,¹⁵ the need to address central PA stenosis and PA hypoplasia was 98% in their patients who underwent the RV-PA Norwood procedure. We hypothesized originally that the 6-mm RV-PA conduit would result in improved central PA growth and reduced PA intervention compared with the 5-mm conduit. This study reports the differences in PA growth between 2 sizes of RV-PA conduit. Use of the 6-mm RV-PA conduit is associated with larger central PA size and had only 1 PA intervention. The PA index in group 1 was not statistically larger than group 2, possibly because of poststenotic dilation caused by the narrow central PA in group 2. However, the higher pulmonary blood flow in group 1 may have resulted in better development of the central and peripheral PA vessels, and may have led to a lower transpulmonary gradient. The larger central PA in group 1 decreased the need for PA augmentation before and during the Glenn operation.

TABLE 2. Pre-Glenn cardiac catheterization data and Glenn operative data at stage 2

	6-mm RV-PA conduit (n = 7)	5-mm RV-PA conduit (n = 8)	P value
Cardiac catheterization data			
Qp/Qs ratio	1.77 ± 0.88	1.06 ± 0.39	.05
Transpulmonary gradient, mm Hg	4.2 ± 1.9	7.8 ± 3.1	.036
Ventricular end diastolic pressure, mm Hg	9.4 ± 2.9	10.5 ± 4.1	.60
Coronary perfusion pressure, mm Hg	39.8 ± 2.9	33 ± 6.3	.026
Minimum size of the central PA, mm	4.5 ± 1.2	2.5 ± 1.0	.009
RPA, mm	8.7 ± 2.7	6.8 ± 2.2	.19
LPA, mm	8.1 ± 3.0	6.2 ± 3.1	.30
Nakata index, mm ² /m ²	416.6 ± 175.4	349.2 ± 276.5	.63
Shunt stenosis (PA site)	14.2% (1/7)	25% (2/8)	.61
Operative data at Glenn			
Age at Glenn, d	194 (104-693)	111 (72-172)	.028
Weight at Glenn, kg	5.2 ± 1.7	4.9 ± 0.3	.78
Off-pump Glenn	100% (7/7)	0% (0/8)	.0002

RV-PA, Right ventricle–pulmonary artery conduit; Qp/Qs, pulmonary-to-systemic flow ratio; PA, pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery.

Infants having the 5-mm RV-PA conduit were referred for Glenn surgery, on average, more than 2 months earlier than those in group 1. Moreover, group 1 patients were better suited for BCPS without cardiopulmonary bypass. Major evolution of the RV-PA conduit was in the site of the right ventriculotomy, which moved cephalad and leftward as our experience evolved between 2002 and 2010. Era effect probably plays a role; however, the anatomy of the PA is a more important determinant not to use cardiopulmonary bypass. A recent report from our institution¹⁸ showed that BCPS without the use of cardiopulmonary bypass can be performed safely and with low mortality and less blood product usage in select infants with HLHS.

Recent advances have led to improved operative outcomes in HLHS and its variants, and we hope this study will advance the surgical strategies for selection of optimal shunt sizes as to decrease PA intervention rate and favor the use of off-pump BCPS. The differences in PA growth might affect long-term Fontan outcomes.

Limitations

The limitations of this report include its retrospective observational nature and relatively small numbers. This study is not powered to evaluate long-term outcome for PA size and ventricular function between comparison groups. The ongoing randomized trial, ideally, will resolve many of these questions and avoid many of the limitations associated with retrospective studies.

CONCLUSIONS

Satisfactory 30-day survival and interstage mortality after stage 1 reconstruction can be achieved with a 6-mm of RV-PA conduit. The 6-mm conduit promotes central PA growth, an improved transpulmonary gradient, and less need for PA intervention and higher success of off-pump BCPS without hospital death.

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