

# The Modified Blalock–Taussig Shunt Versus the Right Ventricle-to-Pulmonary Artery Conduit for the Norwood Procedure

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**Abstract** The initial Norwood procedure remains the highest risk operation for the staged repair of univentricular congenital malformations with associated systemic outflow obstruction. The modified Blalock–Taussig shunt (MBTS) has been implicated as a major cause of not only the operative mortality, but also associated morbidity and interstage attrition. The etiology of these events has often been attributed to the diastolic runoff and “coronary steal” associated with the MBTS, in addition to the delicate balance between systemic and pulmonary blood flow that characterizes all systemic-to-pulmonary artery shunts.

Recently, there has been renewed interest in the right ventricle-to-pulmonary artery conduit as a source of pulmonary blood flow for the Norwood procedure as a potential method for minimizing the negative aspects of the MBTS. The current literature is contradictory, retrospective, and predominantly historically controlled. The Trial of Right Ventricular vs Modified Blalock–Taussig Shunt in Infants with Single Ventricle Defect Undergoing Staged Reconstruction, a randomized controlled clinical trial comparing the two techniques, is ongoing and may provide answers to this controversy.

**Keywords** Hypoplastic left heart syndrome · Norwood procedure · Randomized controlled clinical trial

## The Norwood Procedure with Modified Blalock–Taussig Shunt

Patients with hypoplastic left heart syndrome (HLHS) and other single right ventricle (RV) conditions constitute the highest risk group of patients with congenital cardiovascular malformations; 20 years ago, these conditions were uniformly fatal. The Norwood procedure transformed care of these patients, but hospital mortality remains high, ranging from 7 to 19% in recent series [1, 10, 12, 17, 21, 22]. Postoperative deaths can occur in patients who are clinically gravely ill, or unexpectedly in patients who appear to be making an uneventful recovery [2, 5, 11, 24]. Furthermore, among infants who are discharged from the hospital, 4–15% die between their first and second palliative procedures [2, 24]. In the classic Norwood procedure, pulmonary blood flow is provided by a modified Blalock–Taussig shunt (MBTS), which shunts blood from the innominate or subclavian artery to the pulmonary arteries via a polytetrafluoroethylene (PTFE) tube. The placement of a MBTS results in continuous forward flow from the systemic circulation to the pulmonary circulation throughout both systole and diastole. Because 70–80% of coronary blood flow occurs during diastole, a scenario of “coronary steal” may result as diastolic retrograde flow occurs in both the coronary arteries and descending aorta [9, 15]. Our group has previously demonstrated that coronary arterial flow and oxygen delivery both at rest and after administration of

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**Table 1** Coronary blood flow and oxygen delivery after anatomic or single ventricle repairs for congenital cardiovascular malformations

	Group I (anatomic repair)	Group II (Norwood)	<i>p</i>
Resting coronary flow (ml/min/g)	18 ± 0.2	1.0 ± 0.3	0.003
Coronary flow, after adenosine (ml/min/g)	2.6 ± 0.5	1.5 ± 0.7	0.02
Resting O <sub>2</sub> delivery (ml/min/100 g)	28.9 ± 4.42	16.1 ± 4.2	0.02
O <sub>2</sub> delivery after adenosine (ml/min/100 g)	42.3 ± 5.8	25.5 ± 8.1	0.02

adenosine are significantly decreased in patients after the Norwood procedure compared to patients after anatomic repair of a congenital cardiovascular malformation (Table 1) [6]. Although the exact cause of death in both the hospitalized and the discharged patients is frequently unknown, coronary arterial insufficiency secondary to the diastolic runoff that occurs with a MBTS may play an important role [2, 5, 11].

### The Norwood Procedure with Right Ventricle-to-Pulmonary Artery Conduit

Recently, there has been a renewed interest in the right ventricle-to-pulmonary artery conduit (RVPAC) to supply pulmonary artery blood flow for the Norwood procedure as initially described by Dr. Norwood in some patients early in his experience [14]. A few small, nonrandomized studies have reported short-term improvements in survival with a RVPAC rather than the standard MBTS [10, 12, 17, 21]. The RVPAC has the theoretical advantage of eliminating the aortic diastolic runoff and coronary arterial steal. Our group has previously shown that based on Doppler echocardiography, the typical continuous flow associated with MBTS is not seen with the RVPAC. In addition, the Doppler tracings of the descending aorta demonstrate normal flow patterns with the RVPAC in contrast to the retrograde diastolic flow seen with MBTS [15]. The main theoretical disadvantage of the RVPAC is the need to perform a ventriculotomy, with the potential detrimental effects on ventricular function and arrhythmia incidence.

Other recent studies have shown no improvement in hospital survival comparing the two sources of pulmonary blood flow [1, 10, 22]. Aside from the consistent finding of better historical results with the traditional MBTS at institutions finding no improvement in hospital survival with the RVPAC, there may be other important differences in management between centers to explain the differing results. Sano and associates [21] reported an 89% hospital survival with the RVPAC

compared to 53% with MBTS. However, the majority of the patients in this series undergoing the Norwood with the MBTS received a 4-mm shunt, a size considered too large by most centers with better success utilizing the MBTS. In addition, as is true with most of these studies, the patients are nonrandomized and historically controlled. Pizarro and colleagues [17] demonstrated an improvement in hospital survival from 70% with the MBTS to 92% with the RVPAC. However, again, the majority of MBTS patients received a 4-mm shunt and the controls were historical. Another nonrandomized, historically controlled study by Mair et al. [12] showed an improvement in survival from 72% to 93% with the RVPAC. This group utilized 3.0- to 3.5-mm PTFE grafts for the MBTS. However, the  $Q_p:Q_s$  at the time of the pre-stage II catheterization was 1.55:1 compared to 0.86:1 for the RVPAC, indicating a much higher pulmonary blood flow in the MBTS patients. A  $Q_p:Q_s$  of 1.55:1 has been found to be too generous at the time of the Norwood procedure, much less at the time of the stage II procedure when the shunt has become relatively smaller due to somatic growth. A  $Q_p:Q_s$  closer to 1:1 after the Norwood procedure will result in better systemic flow and has been calculated to be optimal by both mathematical and computer modeling, as well as clinic experience [3, 13].

These results are in contrast to publications from other groups, including Mahle and associates [10], who demonstrated no difference in hospital survival between the MBTS and the RVPAC (81 vs 81%) in a retrospective, historically controlled study. This group utilized MBTS sizes ranging from 2.5 to 4 mm constructed from PTFE ( $n = 2$ ) or saphenous vein grafts ( $n = 20$ ). In this study, the mean  $Q_p:Q_s$  was identical at 0.9:1 prior to stage II for the two groups, suggesting that equivalent pulmonary blood flow may result in equivalent survivals. Similarly, Azakie et al. [1] demonstrated no difference in hospital survival (90 vs 90%) in another retrospective, historically controlled study utilizing 3.5-mm PTFE grafts for the MBTS. In the single study using contemporary but nonrandomized controls, Tabbutt and colleagues [22]

again showed no difference in hospital survival using predominantly a 3.5-mm MBTS (86%) compared to the RVPAC (84%).

In addition to the conflicting data regarding hospital survival, there are many other outcomes for which we have either no data or conflicting information, including the effect of the two shunts on long-term survival, RV function, tricuspid valve function, arrhythmia occurrence, interstage mortality, stage II timing and outcomes, and pulmonary artery growth. Another question that remains to be addressed is the effect of diastolic runoff on other end organ function, such as the brain. The limited data that are available include a study by Tanoue and colleagues [23], which found that following the stage II operation, patients with a RVPAC had worse right ventricular contractility compared to the cohort with a MBTS. Both Tabbutt et al. [22] and Pizarro et al. [18] found a significant decrease in interstage mortality with the RVPAC. However, Tabbutt and associates [22] also noted a significantly higher need for shunt reintervention in the RVPAC group. Several groups have reported no difference in the timing of the stage II procedure; however, Januszewska et al. [8] reported that the aortic and superior vena cava oxygen saturations were significantly lower in the RVPAC group. Interestingly, despite lower oxygen saturations in the RVPAC cohort, Januszewska et al. also noted significantly better pulmonary artery growth prior to stage II, as did Tabbutt et al. Conversely, Sano and colleagues [20] reported 26 of 41 patients (63%) undergoing a stage II operation after a Norwood procedure with a RVPAC required concomitant intervention for pulmonary artery stenosis, including 9 instrumental manual dilations and 17 reconstructions.

### The Single Ventricle Reconstruction Trial

In an effort to address the many unknowns regarding the role of the MBTS and the RVPAC in the management of the patient requiring a Norwood procedure, the Trial of Right Ventricular vs Modified Blalock-Taussig Shunt in Infants with Single Ventricle Defect Undergoing Staged Reconstruction [Single Ventricle Reconstruction (SVR) trial was undertaken. This multiinstitutional, randomized clinical trial is being performed under the auspices of the Pediatric Heart Network (PHN; <http://www.pediatricheartnetwork.com>) with funding from the National Heart, Lung, and Blood Institute, National Institutes of Health, Department of Health and Human Services. Participating centers include the seven core institutions

**Table 2** Core institutions and auxiliary sites of the Pediatric Heart Network

Core institutions
Children's Hospital Boston, Boston, MA
Children's Hospital of New York, New York, NY
Children's Hospital of Philadelphia, Philadelphia, PA
Duke University Medical Center Consortium, Durham, NC/Winston Salem, NC/Greenville, NC
Hospital for Sick Children, Toronto, Ontario, Canada
Medical University of South Carolina, Charleston, SC
Primary Children's Medical Center, Salt Lake City, UT
Auxiliary sites
Children's Hospital of Los Angeles, Los Angeles, CA
Children's Hospital of Wisconsin, Milwaukee, WI
Cincinnati Children's Hospital, Cincinnati, OH
Congenital Heart Institute of Florida, Tampa/St. Petersburg, FL
Emory University, Atlanta, GA
University of Michigan, Ann Arbor, MI

of the PHN and six auxiliary sites (Table 2). New England Research Institutes (Watertown, MA) serves as the network data coordinating center.

The SVR trial is a randomized controlled clinical trial comparing the outcomes of patients with single right ventricle malformations undergoing a Norwood procedure. The patients are randomized to receive either a MBTS or RVPAC. The primary aim of the SVR trial is to compare the incidence of death or transplantation at 12 months of patients undergoing a Norwood procedure with either a MBTS or RVPAC. Secondary aims are to evaluate the hospital and intensive care unit course at the time of the Norwood and stage II procedures, the RV function, the pulmonary artery growth, unintended cardiovascular interventions, and neurodevelopmental outcome at 14 months. The target sample size is 233 patients per treatment arm, for a total of 466 patients. Based on center volumes from previous years, enrollment, which began in May 2005, is expected to continue until summer 2007 [16].

### Comments

The SVR trial is of singular importance not only to help in determining the roles of the MBTS and the RVPAC in the care of the patient undergoing a Norwood procedure, but also to demonstrate that as a specialty, we believe in and are committed to evidence-based medicine. It is sobering to realize that prior to the SVR trial there has never been a large randomized controlled clinical trial in congenital heart surgery comparing two surgical techniques. Until now, the gold standard has been to conceptualize a new

technique or operation, try it on a cohort of patients, and, if it seems to be better compared to a group of historical controls, publish it as fact. Aside from the bias to only publish positive results, it has also clearly been shown that historically controlled trials inappropriately favor the new intervention [4, 7, 19].

Although useful to a point, this approach is no longer acceptable. Phenomenal strides have been made in pediatric oncology directly as a consequence of the fact that essentially every child with a malignancy is enrolled in a protocol. Although it is tempting to state that it is difficult to perform randomized trials in congenital heart disease due to the fact that no one center sees enough of a given malformation, one must remember that congenital heart disease is more prevalent than childhood cancer.

As the health care professionals providing care for our patients, we should be on the forefront, championing the need for evidence-based medicine in optimizing the management of congenital heart disease. External pressures are mounting from government (the New York State Cardiac Surgery Reporting System) and from third-party payers (the Leapfrog Group) for transparency and consistent results. Compounded with the public and the media interest (one might even say frenzy) in the topics of medical errors, the 80-hour workweek, the FDA, the Bristol Affair, and Vioxx (Merck, Whitehouse Station, NJ, USA), if we do not take the lead, someone else surely will.

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