Primary Versus Staged Repair in Neonates With Pulmonary Atresia and Ventricular Septal Defect

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Background. **The 2 surgical strategies for neonates with ductal-dependent pulmonary atresia and ventricular septal defect are primary biventricular repair (BVR) or initial palliation with a modified Blalock-Taussig shunt (BTS) followed by second stage repair. In this study, we report the combined outcomes from 2 hospitals using different strategies.**

Methods. **Between 2004 and 2017, 66 neonates underwent surgery with palliative shunts (BTS group: n = 30, 45.5%) or primary biventricular repair (pBVR group: n = 36, 54.5%). The 2 groups were similar in age, body weight, and Nakata index scores. The overall mean follow-up duration was 7.51 ± 4.35 years, and early and late results were compared between the groups.**

Results. **The 10-year overall survival was 84.8% (94.4% for pBVR vs 75.7% for BTS, $P = .032$). The BTS group had 2 early and 6 interstage mortalities, and the pBVR group had no early and 2 late mortalities. In the BTS group, the Nakata index score significantly increased during the interstage period ($P < .001$). In univariable analysis, genetic or extracardiac anomalies were a risk factor for mortality (hazard ratio, 5.56; $P = .038$). After achieving BVR, the pBVR group underwent significantly more frequent right ventricle outflow tract reinterventions ($P < .001$) at a much earlier period ($P = .017$) compared with the BTS group.**

Conclusions. **In neonates with ductal-dependent pulmonary atresia and ventricular septal defect, the primary BVR approach provides an excellent survival rate, but the burden of right ventricle outflow tract reintervention is heavy. The staged approach with BTS promotes pulmonary artery growth, but hospital and interstage mortality are significant. Genetic and extracardiac anomalies are significant risk factors for mortality.**

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Pulmonary atresia with ventricular septal defect (PA VSD) is a rare congenital cardiac anomaly that affects between 4.2 and 10 per 100,000 live births.[1-3] Although the right ventricle is usually well developed, the pulmonary arteries (PAs) are extremely heterogeneous in anatomy with regard to size, shape, and collateral vessels. In patients with reasonably sized PAs, the patent ductus arteriosus acts as the primary source of pulmonary blood flow. On the other hand, if the PAs are absent or small, the pulmonary circulation is maintained through the multiple major aortopulmonary collateral arteries (MAPCAs). The ductal-dependent type of PA VSD is generally considered to be less complicated, as unifocalization of the MAPCAs is not required.[4,5] Nevertheless, these patients require early surgical intervention in the neonatal period to create a more reliable blood supply to the lungs.

The traditional surgical option for neonates with ductal-dependent PA VSD has been to perform initial palliation with a Blalock-Taussig shunt (BTS) followed by a second stage biventricular repair (BVR) after the patient had grown. As operative techniques and perioperative management have advanced, however, early primary BVR has been adopted, similar to the treatment paradigm for tetralogy of Fallot.[4] Several studies have highlighted the advantages and shortcomings of each strategy.[4-8] Because of the low prevalence, however, it is not easy to collect a large cohort of PA VSD patients to perform an adequate comparison of these 2 surgical options.

Seoul National University Children's Hospital (SNUCH) and Sejong General Hospital (SJGH) are the leading and longest-running congenital cardiac centers in South Korea. Although these 2 centers have worked closely for over 40 years, they have different approaches to treating ductal-dependent PA VSD. Surgeons at SNUCH prefer to perform a staged repair with BTS,

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Abbreviations

BTS	=	Blalock-Taussig shunt
BVR	=	biventricular repair
MAPCA	=	major aortopulmonary collateral artery
PA	=	pulmonary artery
PA VSD	=	pulmonary atresia with ventricular septal defect
pBVR	=	primary biventricular repair
RVOT	=	right ventricle outflow tract
SJGH	=	Sejong General Hospital
SNUCH	=	Seoul National University Children's Hospital
VACTERL	=	vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities

whereas surgeons at SJGH almost always perform primary BVR. In this study, we analyze the combined outcomes of these 2 hospitals to determine which surgical strategy has the best short- and long-term results.

Material and Methods

Patient Selection and Data Collection

Between 2004 and 2017, 36 and 30 neonates with ductal-dependent PA VSD underwent surgery at SNUCH (54.5%) and SJGH (45.5%), respectively. These 66 patients were combined and classified into either the BTS group (n = 30, 45.5%) or the pBVR group (n = 36, 54.5%) according to the surgical strategy. Patients with significant MAPCAs were excluded. The diameters of the left and right PAs immediately proximal to the origin of their respective upper lobe branches were measured by both echocardiography and computed tomography.[9] The Nakata index was calculated using the larger measurements by dividing the sum of the cross-sectional area of the 2 PAs by the body surface area of the patient.[10] Other medical records, including perioperative hospital course, operative reports, and follow-up studies, were also reviewed and analyzed retrospectively. The overall mean follow-up duration was 7.51 ± 4.35 years, and early and late results were compared between the BTS group and the pBVR group. The protocol for this study was approved by the ethics committees of both hospitals.

Two Hospitals With Different Strategies

Of the 36 patients at SNUCH, most underwent staged repair with a BTS as initial palliation (80.6%, 29 out of 36). Primary BVR was performed in 6 patients who had well-developed branch pulmonary arteries without stenosis or hypoplasia. One patient with VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) association had to undergo several surgical corrections during infancy and was considered to be at high risk for interstage mortality after BTS. Therefore, even with small pulmonary arteries (Nakata index 87.5), our multidisciplinary team decided to perform a primary BVR.

A total of 93% of the patients (28 out of 30) at SJGH underwent primary BVR. BTS was performed in only 2 patients. One patient had a marginally small left ventricle, and the other had severe pulmonary artery hypoplasia (Nakata index 41.7).

Echocardiography was the primary means of evaluation during follow-up in both hospitals. Cardiac catheterization, computed tomography, or cardiac magnetic resonance imaging was also performed if necessary. Although the general principle of reintervention was similar between the 2 hospitals, doctors at SJGH tended to utilize percutaneous intervention more aggressively.

Statistical Analysis

The data are presented as median with interquartile range, mean with SD, or frequencies, as appropriate. Qualitative variables (n with percentages) were analyzed with the χ^2 test. Continuous variables were compared with *t* tests, Mann-Whitney U tests, or Wilcoxon matched-pairs tests, as appropriate. The Z-score was calculated using the Detroit *z*-score package.[11] Probabilities of time-dependent outcomes (death and right ventricle outflow tract [RVOT] reintervention) were estimated with the Kaplan-Meier method and compared by log-rank analysis. A *P*-value less than .05 was considered statistically significant. All statistical analyses were performed with SPSS Statistics, version 20.0.0.1 (IBM, Armonk, NY).

Results

Patient Characteristics and Operative Details

The mean age at the time of the first operation was 18.05 ± 7.71 days, and the mean body weight was 3.23 ± 0.46 kg (Table 1). There was no difference in birth weight, operation body weight, corrected gestational age, or Nakata index between the BTS and the pBVR groups. Seven patients (10.6%) were born prematurely (<37 weeks). Genetic or extracardiac anomalies were present in 10 patients, including DiGeorge syndrome (n = 3), Miller-Dieker syndrome, imperforate anus (n = 2), VACTERL association, Alagille syndrome, microtia, and meningocele.

In the BTS group, concomitant angioplasty was performed in 17 patients (56.7%). Unplanned reoperations during the hospital stay included extracorporeal membrane oxygenation support (n = 2), additional right modified BTS procedure, and shunt revision (n = 2). During postoperative intensive care unit care, 4 patients suffered cardiac arrest, and 2 of them did not survive. The mean interval between the first- and second-stage operations was 0.8 ± 0.46 years in the BTS group. During this period, 6 patients died due to cardiac arrest. However, since 2012, there has been only 1 interstage mortality among 16 patients. This patient had multiple anomalies consistent with the VACTERL association. Five months after the BTS procedure, she was admitted to the general

Table 1. Demographic Data and Morphologic Features Before the First Operation

Characteristics	BTS Group (n = 30)	pBVR Group (n = 36)	*P* Value
Male sex	14 (46.7)	17 (47.2)	.964
Prematurity (<37 wk)	3 (10.0)	4 (11.1)	.960
Birth weight, kg	2.9 ± 0.41	2.9 ± 0.45	.981
Genetic/extracardiac anomalies	6 (20)	4 (11.1)	.492
First operation			
Operation age, d	19.63 ± 9.52	16.72 ± 5.6	.360
Corrected gestational age, d	292.63 ± 11.33	289.58 ± 9.57	.240
Operation body weight, kg	3.3 ± 0.41	3.17 ± 0.49	.244
Nakata index	119.97 ± 48.92	117.22 ± 34.93	.794
PA stenosis	20 (68.97)	19 (54.29)	.231
PA hypoplasia	12 (41.38)	10 (28.57)	.283

Values are presented as mean ± SD or n (%).

BTS, Blalock-Taussig shunt; pBVR, primary biventricular repair; PA, pulmonary artery.

surgery department to undergo a Peña operation and colostomy repair for ileal atresia. Fluid balance was not adequately managed during bowel preparation, and the patient went into cardiac arrest the night before surgery. Twenty-two patients in the BTS group successfully underwent a second-stage operation, with a mean bodyweight of 8.37 ± 1.92 kg (Table 2). The mean Nakata index score was significantly greater than that at the time of the previous operation (119.97 ± 48.92 vs 188.65 ± 99.8, $P < .001$). Valved conduits were used in all but 4 patients. There were no early or late mortalities.

Meanwhile, the pBVR group underwent total correction using RVOT patch and pericardial roll in 23 and 13 patients, respectively. Thirteen patients (36.1%) underwent concomitant angioplasty, and 5 patients (13.9%) had a delayed closure of the sternum. There were 2 unplanned reoperations for residual VSD leak and severe RPA stenosis. Early and late mortalities in the pBVR group were 0 and 2 patients, respectively.

Time-Related Survival After Surgery

Overall survival at 1 and 10 years after the first operation was 87.9% and 84.8%, respectively (Figure 1). Eight out of 10 mortalities occurred within 6 months after the surgery, and none occurred after 2 years. The survival rates at 1 month, 1 year, and 10 years for each treatment group were 93.3%, 80%, and 75.7% for the BTS vs 100%, 94.4%, and 94.4% for the pBVR group, respectively ($P = .082$) (Figure 2). In univariable analysis, the presence of genetic or extracardiac anomalies was a risk factor for mortality (hazard ratio, 5.56 (95% confidence interval 1.1%-28.02%); $P = .038$).

Table 2. Operative Characteristics of Patients Who Underwent Second-Stage BVR in the BTS Group

Second-Stage BVR	BTS Group (n = 22)
Operation age, y	0.86 ± 0.46
Interstage period, y	0.8 ± 0.46
Operation body weight, kg	8.37 ± 1.92
Nakata index	188.65 ± 99.8
RVOT reconstruction method	
Trans-annular patch	4
12-mm valved conduit	8
14-mm valved conduit	8
16-mm valved conduit	2

Values are presented as mean ± SD or number of patients.

BTS, Blalock-Taussig shunt; BVR, biventricular repair; RVOT, right ventricular outflow tract.

Reintervention for RVOT After Biventricular Repair

A total of 58 patients were followed up for a mean duration of 6.98 ± 4.2 years after achieving BVR (Table 3). Overall, compared with the BTS group, the pBVR group underwent significantly more frequent RVOT reinterventions at a much earlier period. In the BTS group, the percentages of patients free from RVOT reintervention at 1 year, 2 years, and 5 years after BVR were 90.5%, 84.8%, and 40.0%, respectively (Figures 3A, 3B). During a mean follow-up period of 5.07 ± 3.75 years, 6 percutaneous

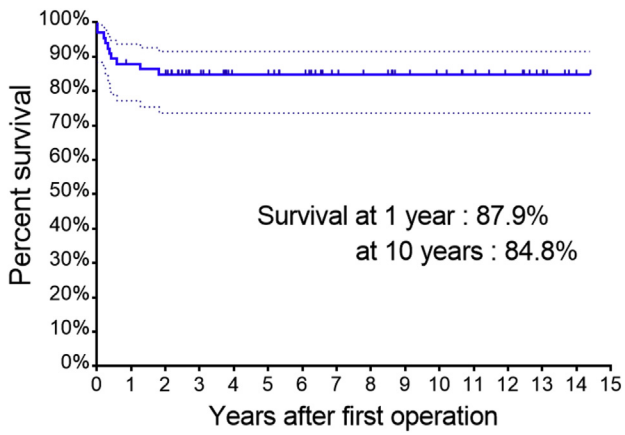

Figure 1. Survival for the total cohort of 66 neonates with ductal dependent pulmonary atresia and ventricular septal defect after the first operation.

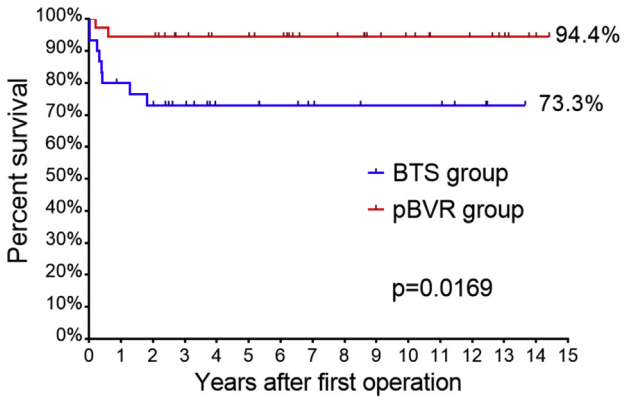

Figure 2. Comparison of survival after first operation: staged repair group (blue line) vs primary repair group (red line). (BTS, Blalock-Taussig shunt; pBVR, primary biventricular repair.)

interventions and 8 reoperations were performed in 10 patients. The shortest interval between the second-stage BVR and reoperation for RVOT was 3.4 years. On the other hand, the BVR group showed significantly less freedom from RVOT reintervention at 1 year, 2 years, and 5 years with values of 42.9%, 28.6%, and 25.0%, respectively. Thirty patients underwent 42 balloon angioplasties, 14 stent insertions, and 32 reoperations, and 53.3% of the patients had more than 3 RVOT reinterventions. The median interval between BVR and the first RVOT reintervention was 0.62 years (range, 0.23-13.75 years).

Prognosis of the 'Non-traditionally' Treated Patients of Each Hospital

Primary BVR was performed in 7 patients at SNUCH. There were 2 catheter-based interventions and 6 reoperations during a mean follow-up period of 6.96 ± 3.35 years (1.14 RVOT reintervention event per patient). Mean interval between the initial operation and first RVOT reintervention was 2.36 ± 2.83 years. There was no statistical significance between the total pBVR group and this subgroup. Of the 2 BTS patients from SJH, 1 underwent BVR at the age of 1.7 years. This patient had 1 reoperation for pulmonary valve replacement and is currently alive and well after 13.7 years. The other BTS patient had multiple anomalies, including Alagille syndrome. He died of infection before completing BVR.

Comment

PA VSD neonates without MAPCAs usually have to rely on the unpredictable ductus arteriosus to supply the lungs, which means that there is always a risk of severe life-threatening hypoxia or heart failure. Therefore, early surgical intervention to create a stable and controlled pulmonary blood supply is necessary. Currently, the 2 main surgical options are a 2-stage approach with initial palliation with a BTS and delayed BVR or a single-stage approach with neonatal primary BVR. Because of the rarity and diversity of this malformation, there are only a handful of reports that have presented a direct comparison between the 2 surgical options in ductal-dependent PA VSD patients. Our colleagues from SJGH previously presented the results of both strategies in the early era of primary BVR.[6] Compared with the shunt palliation group, the primary repair group had fewer mortalities but suffered from more frequent reintervention and earlier reoperation of the RVOT. More recently, Alsoufi and associates[12] have reported the largest series to date with 86 neonates. In their analysis, patients who underwent primary BVR showed a trend towards better survival but worse freedom from RVOT reoperation. Genetic and extracardiac anomalies were identified as risk factors for mortality.

In this study, we compared the short and mid-term outcome of single-stage and 2-stage strategies in the modern era. Our results are consistent with the 2 aforementioned studies in terms of survival and reoperation rate.

Multiple studies have highlighted the pros and cons of BTS in patients with various cardiac anomalies.[7,13-15] Being able to avoid or reduce the use of cardiopulmonary bypass during the fragile neonatal period is a significant advantage of the BTS procedure. Furthermore, the heart and PAs increase in size, which makes it much easier for the surgeon to avoid PA kinking or stenosis during the second-stage BVR. In our study, the BTS group showed a more than 40% increase in the Nakata index score during

Table 3. Data of RVOT Reintervention After Achieving BVR is Compared Between the 2 Groups With Different Surgical Approach

Characteristics	BTS Group (n = 22)	pBVR Group (n = 36)	*P* Value
Follow-up period, y	5.07 ± 3.75	7.51 ± 4.35	.023
RVOT re-intervention (event per patient)	14 (0.64 ± 0.85)	88 (2.44 ± 2.02)	<.001
Percutaneous intervention	6	56	
Reoperation	8	32	
Time to first RVOT re-intervention, y	3.72 ± 2.62	1.93 ± 3.32	.017
Time to first percutaneous intervention, y	3.06 ± 3.02	1.01 ± 1.86	
Time to first reoperation, y	5.46 ± 2.25	3.85 ± 4.24	

Values are presented as mean ± SD.

BTS, Blalock-Taussig shunt; BVR, biventricular repair; pBVR, primary biventricular repair; RVOT, right ventricular outflow tract.

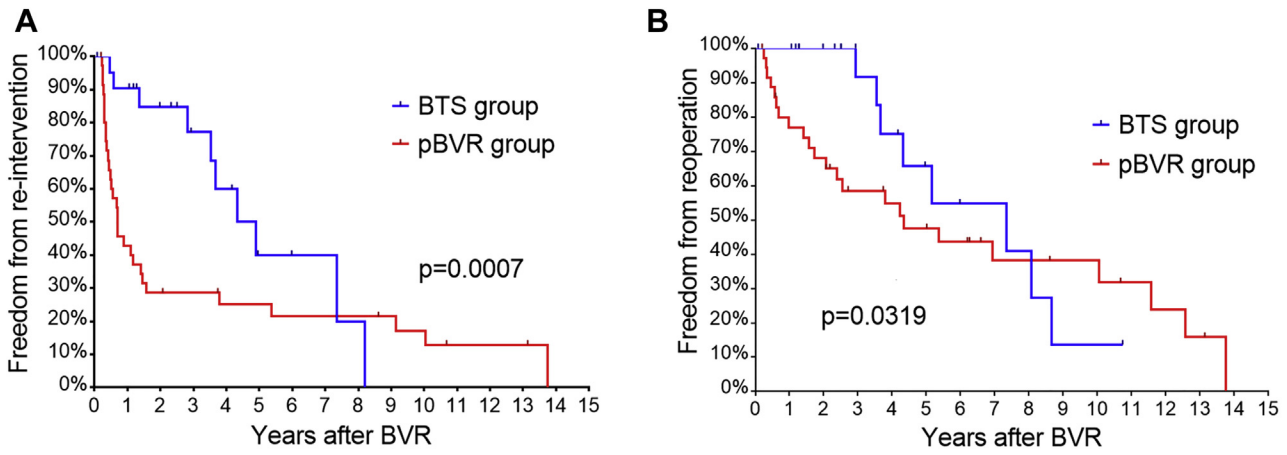

Figure 3. Freedom from (A) all reintervention (percutaneous intervention and reoperation) and (B) reoperation for the right ventricle outflow tract after achieving biventricular repair. (BTS, Blalock-Taussig shunt; pBVR, primary biventricular repair.)

the interstage period. This finding coincides with other reports suggesting that BTS promotes PA growth.[4,16] This, we believe, is the reason the BTS group underwent far less percutaneous intervention than the pBVR group after the correction was completed. Along with the intrapericardial structures, the chest cavity is also large enough to fit a reasonably sized valved conduit. In our study, valved conduits with a diameter of 12 mm or larger were used in 18 out of 22 patients (81.8%) during the second stage BVR. Meanwhile, in the pBVR group, a valved conduit could not be used during the first operation because of the small patient size. Although these small valved conduits have a short life cycle, they can protect the right ventricle from early exposure to pulmonary regurgitation. Overall, the BVR in the BTS group seemed to have a "better quality" than that in the pBVR group in that patients underwent less percutaneous intervention (6 vs 56), underwent less reoperation for the RVOT (8 vs 32), and had a longer interval between the BVR and the next operation (5.43 vs 2.06 years).

On the other hand, significant in-hospital and interstage mortality after BTS ranging anywhere between 5.1% and 14% is a well-known issue.[13-15] The superior overall survival of the primary BVR approach is consistent across multiple reports, as was the case in our study.[4,6,12] This tendency especially stands out when patients have a genetic or extracardiac anomaly. Although the prevalence of associated anomalies was not as high as Alsoufi and coworkers[12] reported (15.2% vs 40%), it was still a significant risk factor for mortality. Among the 10 patients who had a genetic or extracardiac anomaly, 6 patients underwent BTS, and 4 of them did not survive to the second stage. Meanwhile, only 1 out of 4 patients with combined anomalies died in the pBVR group. In addition to their anatomical differences and relation to prematurity and low body weight, the fact that some of these patients have to undergo noncardiac-related medical or surgical procedures during the interstage period increases their risk of mortality. Furthermore, as we learned from our experience, the medical staff in other departments do not always have an adequate understanding of the delicate hemodynamics of a palliative state patient, which can lead to disastrous results. After losing a patient during the preparation for a noncardiac surgery, we tightened our policy to (1) postpone all noncritical extracardiac surgical procedures until BVR is achieved and (2) maintain stronger interdepartmental communication and cooperation.

In recent years, many centers have attempted to overcome the risks in the vulnerable interstage period by introducing a more stringent follow-up protocol.[12,17,18] In 2013, SNUCH also implemented an interstage-care program that provides home monitoring and phone counseling for patients with BTS. Upon discharge from the hospital, the parents are instructed to monitor the patient's oxygen saturation at home and call a dedicated line if low oxygen saturation, persistent fever, vomiting, diarrhea, reduced oral intake, or any other signs of distress appear. An experienced physician's assistant discusses the child's condition with the parents and advises them to seek early medical care if necessary. This program, we believe, is one of the main reasons we were able to reduce interstage mortality in recent years.

As our understanding of congenital cardiac disease evolves, the focus of treatment moves beyond simple survival and into improving the long-term results and quality of life of the patient. When choosing a surgical option for a PA VSD patient, we have to consider not only the characteristics and anatomy of the patient but also the context of the medical and socioeconomic situation. Our results suggest that compared to a primary BVR approach, staged repair with BTS promotes PA growth and thus reduces the frequency of RVOT-related reintervention in the long term. This is a substantial advantage, as repeated intervention and reoperation have a considerable impact on the patient's life as well as the lives of their family members. However, until BVR is achieved, patients in a palliative state require delicate care not only during their hospital stay but also after returning home. Good family support, frequent

counseling, and ready access to a pediatric cardiac center are crucial elements to avoid tragic outcomes. On the other hand, the primary BVR approach shows excellent survival in both the short and long term. Therefore, it might be safer to perform primary BVR in high-risk patients, especially those with genetic or extracardiac anomalies—although collaboration with a cardiologist with adequate skill and experience is needed.

Study Limitations

As a retrospective observational study, this study has inherent limitations, such as selection bias and the lack of randomization. Although both centers share a similar paradigm, the positive attitude of SJGH towards percutaneous intervention clearly had an impact on the frequency of RVOT intervention in the pBVR group. Even with the combined results from 2 centers, the cohort was small, precluding more sophisticated statistical analyses and further identification of risk factors.

Conclusions

There is no easy answer when choosing the best surgical strategy in ductal-dependent PA VSD neonates. The primary BVR approach provides an excellent survival rate but leads to frequent RVOT intervention and reoperation. On the other hand, the staged approach with BTS promotes pulmonary artery growth and thereby improves the quality of the second stage BVR. Nevertheless, the risk of hospital and interstage mortality is still significant. The choice of operation should be individualized and based on the situation of each patient in each institution. Genetic or extracardiac anomalies are a significant risk factor for mortality, and primary BVR might provide a better chance of survival for patients with these conditions.

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