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The Recent Surgical Result of Total Anomalous Pulmonary Venous Return

So-Ick Jang, MD¹, Jin-Young Song, MD¹, Soo-Jin Kim, MD¹, Eun Young Choi, MD¹, Woo Sup Shim, MD¹, Chul Lee, MD², Hong-Gook Lim, MD² and Chang Ha Lee, MD²

¹Departments of Pediatrics and ²Cardiac Surgery, Sejong General Hospital, Bucheon, Korea

ABSTRACT

Background and Objectives: Recent surgical results from total anomalous pulmonary venous return (TAPVR) are reported to have improved; however, mortality and morbidity are still high in the univentricles. This review was performed in order to demonstrate surgical results from TAPVR for recent years in a single institute. **Subjects and Methods:** A total of 53 patients with TAPVR underwent surgery between January 2000 and December 2008. Preoperative anatomy and hemodynamics were evaluated by 2-dimensional echocardiogram. We reviewed medical records on preoperative management, age, and body weight at time of surgery, postoperative mortality, and pulmonary venous obstruction. **Results:** The study included 36 biventricle patients and 17 univentricle patients. Median age and weight at time of surgery were 26 days and 3.5 kg, respectively. During the mean follow up period of 48.2 months, a total of 11 deaths and 8 early deaths occurred. Postoperative pulmonary venous stenosis occurred in 8 patients. Among 36 biventricle patients, mortality occurred in 1 patient and postoperative pulmonary venous stenosis occurred in 6 patients. No statistical significances were observed between postoperative pulmonary venous stenosis and other factors. Among 17 univentricle patients, mortality was 10, including 7 early deaths, and postoperative pulmonary venous stenosis occurred in 2 patients. Combined palliations were modified Blalock-Taussig (BT) shunts in 7, pulmonary artery banding (PAB) in 3, bidirectional cavopulmonary shunt in 5, and the Fontan procedure in 1. The highest mortality was recorded in patients with a modified BT shunt (85.7%) and next in patients with PAB (66.6%). Mortality was not related to anatomy of TAPVR, nor preoperative obstruction, but with significant age and weight at time of surgery. **Conclusion:** Despite improvement in operative results from TAPVR, surgery in univentricles is highly risky, especially with low weight and early age, with concomitant palliation. (*Korean Circ J* 2010;40:31-35)

KEY WORDS: Pulmonary vein; Anomaly; Univentricular heart.

Introduction

Recent surgical results from total anomalous pulmonary venous return (TAPVR) have been reported to be improved due to development of diagnosis and perioperative care.^{1,2)} The survival rate for isolated TAPVR in a biventricular heart is greater than 90%, but in cases of complex TAPVR combined with univentricular

heart or heterotaxy, the survival rate is still reported at less than 60%.²⁻⁵⁾

Chu et al.⁶⁾ reported that operative mortality of TAPVR with biventricular heart was 16.7%, with no deaths since 2000. However, Lee et al.⁷⁾ reported that early postoperative mortality was 43.6% in cases of TAPVR combined with univentricular heart until 2002. Therefore, surgical correction of TAPVR combined with univentricular heart is still expected with high risk mortality. This retrospective review was performed to demonstrate the surgical results of TAPVR between isolated and combined with univentricular heart for recent years in a single institute.

Subjects and Methods

A total of 53 patients with TAPVR underwent surgery at Sejong general hospital from January 2000 to

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Correspondence: So-Ick Jang, MD, Department of Pediatrics, Sejong General Hospital, Sosabon 2-dong, Sosa-gu, Bucheon 422-711, Korea
Tel: 82-32-340-1122, Fax: 82-32-340-1236
E-mail: prelud15@nate.com

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December 2008. Patients were classified with either univentricle or biventricle physiology according to whether their anatomy was considered suitable for biventricle or univentricle repair. Thirty six (67.9%) patients had isolated TAPVR with biventricular heart, and 17 (32.1%) patients had complex TAPVR combined with univentricular heart. Isolated TAPVR was diagnosed if the patient had TAPVR in association with a secundum atrial septal defect (ASD) or a patent ductus arteriosus (PDA), or both.

Preoperative anatomy and hemodynamic was evaluated by 2-dimensional echocardiogram and 3-dimensional chest computed tomography. The obstruction was evaluated with echocardiography (continuous pulmonary venous inflow or accelerated inflow >2 m/s in color Doppler) and/or with chest computed tomography. Early mortality was defined as death that occurred in the first 30 days following the operation. Late mortality was defined as death that occurred more than 30 days after surgery or after discharge from the hospital. We retrospectively reviewed medical records with regard to preoperative management, age, and body weight at operation, postoperative mortality, and pulmonary venous stenosis.

Statistical analysis was performed by Statistical Package for the Social Sciences (SPSS; SPSS Inc, Chicago, IL, USA) 11.5 for windows. MannWhitney test and Fisher's exact test were used for univariable analysis. The study end point was the point of death or December 31, 2008 in survivors.

Results

There were 53 patients of whom 29 were male (54.7%). The median age and weight at time of surgery was 26 days (range; 1-5,948 days) and 3.5 kg (range; 2.0-49.6 kg), respectively. The TPAVR anatomy was supracardiac in 31 cases (58.5%), cardiac in 7 (13.2%), infracardiac in 12 (22.6%), and mixed in 3 (5.7%) (Table 1 and

2). Preoperative pulmonary vein obstruction was high in infracardiac and mixed (13 of 15, 86.7%). During the mean follow up period of 48.17 months, there were a total of 11 deaths (20.8%), and 8 early deaths (15.1%) were found. Postoperative pulmonary venous stenosis occurred in 8 (15.1%) of 47 patients, except for early deaths (Fig. 1).

Biventricular group

Among 36 biventricular patients, there were 18 males and 18 females. Median age and weight at time of surgery were 23.5 days (range; 1-3,085 days) and 3.5 kg (range; 1.9-10.2 kg), respectively. The anatomic subtype with obstruction was described in Table 1.

In the preoperative state, arterial oxygen saturation was $83.8 \pm 6.5\%$, and a mechanical ventilator was applied in 12 (33.3%), intravenous drugs used in 14 (38.9%) with continuous infusion of inotropics, and in 2 with eglandin (prostaglandin E_1). Early mortality occurred in 1 patient (2.8%), and postoperative pulmonary venous stenosis occurred in 6 patients (17.1%). No statistical significances were observed between postoperative pulmonary venous stenosis or other factors.

All 6 postoperative pulmonary stenosis patients survived, 5 in anastomotic sites and 1 in the individual pulmonary vein. Individual stenosis was a required reoperation because of severe ostial narrowing of pulmonary veins after 8 months. We did a wide excision of the stenotic tissue around the pulmonary vein ostia and the sutureless technique using a pericardial patch was done. However, no improvement of pulmonary venous return was noticed, and 20 days later, the second operation was done. Previous patches were excised and the stenotic ostia was opened wide and atrio-pericardia anastomosis was performed from the inside. Postoperative pulmonary venous stenosis was not statistically related with age and weight at operation, anatomic sub-

Table 1. Anatomic subtype with obstruction in biventricular heart

	Number	Obstruction (%)	Early mortality	Late mortality	PV stenosis	Total death
Supracardiac	17	5 (29.4)	0	0	2	0
Cardiac	6	2 (33.3)	0	0	0	0
Infracardiac	11	10 (90.9)	1	0	3	1
Mixed	2	1 (50)	0	0	1	0
Total (%)	36	18 (50)	1 (2.8)	0	6 (17.1)	1 (2.8)

PV stenosis: postoperative pulmonary vein stenosis

Table 2. Anatomic subtype with obstruction in univentricular heart

	Number	Obstruction (%)	Early mortality	Late mortality	PV stenosis	Total death
Supracardiac	14	9 (64.3)	6	3	1	9
Cardiac	1	0	0	0	0	0
Infracardiac	1	1 (100)	1	0	1	0
Mixed	1	1 (100)	0	0	0	1
Total (%)	17	11 (64.7)	7 (41.2)	3	2 (20)	10 (58.8)

PV stenosis: postoperative pulmonary vein stenosis

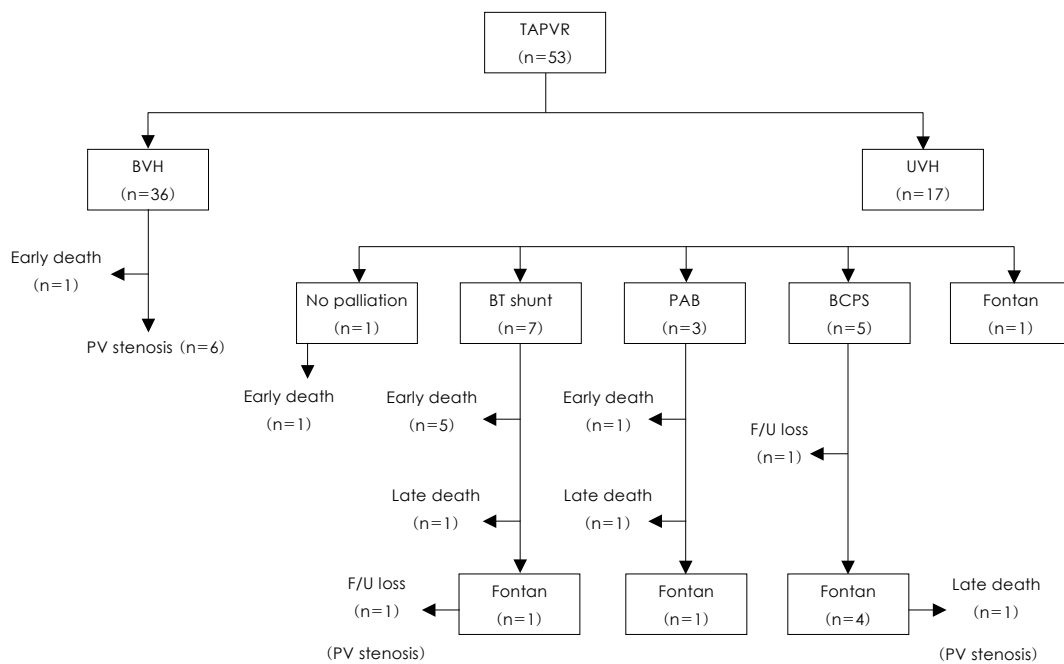


Fig. 1. Intervention and outcome schema for total anomalous pulmonary venous return (TAPVR). BVH: biventricular heart, UVH: univentricular heart, BT shunt: modified Blalock-Taussig shunt, PAB: pulmonary artery banding, BCPS: bidirectional cavopulmonary shunt, F/U: follow up.

types, and preoperative obstruction.

Univentricular group

Among 17 univentricular patients, 11 were male and 6 were female. Median age and weight at time of surgery were 101 days (range; 1-5,948 days) and 3.4 kg (range; 3.1-49.6 kg), respectively. The anatomic subtype with obstruction was described in Table 2. Preoperative pulmonary venous obstruction occurred in 11 patients (64.7%), with pulmonary artery stenosis in 7 patients (41.2%), and pulmonary atresia in 5 patients (29.4%). There were 13 patients (76.5%) with right isomerism.

In the preoperative state, arterial oxygen saturation was $77.2 \pm 6.1\%$, and a mechanical ventilator was applied in 5 (29.4%), intravenous drugs were used in 5 (29.4%) with continuous infusion of inotropics, and in 8 (47.1%) with eglanidin.

Except for 1, all patients underwent total repair of TAPVR with combined palliation using a modified Blalock-Taussig (BT) shunt in 7 (41.2%), and a bidirectional cavopulmonary shunt (BCPS) in 5 (29.4%), pulmonary artery banding (PAB) in 3 (17.7%), and the Fontan procedure in 1 (5.9%). There were 10 deaths (58.8%) with early mortality of 7 (41.2%). Postoperative pulmonary vein stenosis occurred in 2, and 1 patient died. Another patient required reoperation due to anastomotic stenosis with ostial narrowing of bilateral upper pulmonary veins after 9 months. We did a resection of anastomotic fibrotic tissue and wide excision of the stenotic tissue around the pulmonary vein ostia, and no suturing was done because of severe pericardial

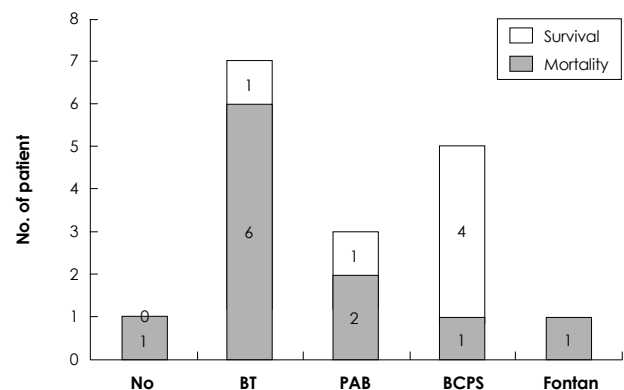


Fig. 2. The relationship between palliative procedure and mortality in univentricular heart. No: no palliation, BT: modified Blalock-Taussig shunt, PAB: pulmonary artery banding, BCPS: bidirectional cavopulmonary shunt.

adhesions. Another case required reoperation for total occlusion of the right pulmonary vein opening and ostial narrowing of the other pulmonary vein after 4 months. For right-sided lesions, stenotic pulmonary vein scar tissue was excised, and then right pericardial wall tissue was anastomosed to the right atrial wall above the left atrial fenestration. For left-sided lesions, repair was conducted through the left atrial cavity and a portion of left atrial tissue was excised around the stenotic and atrio-pericardial anastomosis was done from the inside. However, the patient expired after the Fontan operation with fenestration.

The highest mortality was recorded in patients with modified BT shunt (6 of 7; 85.7%), and the next was in patients with PAB (2 of 3; 66.6%). Of 5 BCPS pa-

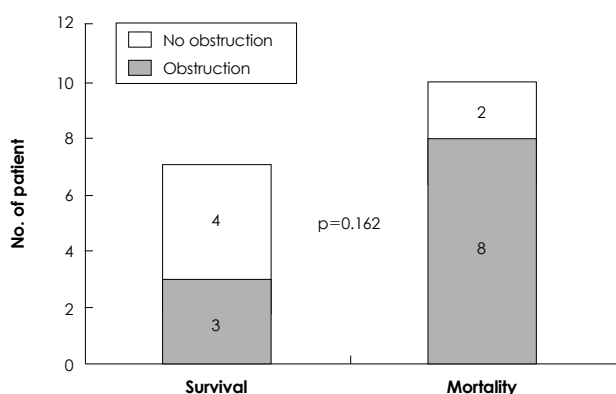


Fig. 3. The relationship between mortality and preoperative pulmonary vein obstruction in univentricular heart. No obstruction: no preoperative pulmonary vein obstruction, Obstruction: preoperative pulmonary vein obstruction.

tients of first palliation, 4 underwent the Fontan procedure, with 1 case of late mortality (Fig. 2). The patient who underwent repair of only TAPVR without palliation died. Mortality was high in patients with modified BT shunt and PAB, but with no statistical significance ($p=0.116$). Postoperative pulmonary stenosis occurred in 8 of 10 mortalities. Univariate analysis showed that mortality was not related to the anatomy of TAPVR, nor preoperative obstruction (Fig. 3), but was significant with regard to age and weight at time of surgery. That is, the age and weight at operation were 42.3 ± 68.5 days and 3.3 ± 1.2 kg, respectively in the mortality group, but $1,066.7 \pm 2,156.3$ days and 12.7 ± 16.3 kg, respectively in the survival group ($p=0.001$). However, preoperative arterial oxygen saturation and support were not statistically related to mortality.

Discussion

Total anomalous pulmonary venous return in biventricular group

Recent reports of surgical correction of TAPVR indicate a general improvement in operative survival. Bando et al.⁸⁾ reported that improvements in surgical technique, as well as preoperative and postoperative management, account for the reduction in mortality and the need for reoperation for most types of TAPVR. However, TAPVR with univentricular heart is still reported to have high mortality, and heterotaxy syndrome is reported as a high risk factor.³⁹⁾

Karamlou et al.¹⁰⁾ reviewed records of 327 children presenting from 1946 to 2005 with TAPVR and reported that overall survival from repair was $65 \pm 6\%$ at 14 years, with a current survival of 97% since 2000. In Korea, Song et al.¹¹⁾ reported the mortality of isolated TAPVR at 8.3% since 1994 and Chu et al.⁶⁾ reported operative mortality at 0% since 2000. This is a marked improvement in surgical results in cases with early age and low body weight at time of surgery. Our study re-

ports that early mortality in isolated TAPVR with biventricular heart is 2.8% (1 of 36), as in other reports. Anatomic subtype and preoperative obstruction, once considered significant risk factors, have been neutralized over time.

However, pulmonary vein stenosis following repair of TAPVR, a significant risk factor for late mortality or reoperation is reported to occur at 0 to 18%.¹²⁻¹⁵⁾ Recently, Chu et al.⁶⁾ reported that postoperative pulmonary vein stenosis occurred at 20.8%, and was 17.1% in this study. However, anastomotic site stenosis is not necessary for surgery, but individual vein stenosis needs aggressive therapy. In this study, we could not find any risk factors for postoperative pulmonary vein stenosis.

Total anomalous pulmonary venous return in univentricular group

Early mortality of TAPVR combined with a univentricular heart is still reported as very high, and Gaynor et al.⁴⁾ reported that survival for patients who underwent TAPVR surgery combined with univentricular heart was 54% at 6 months of age, and 44% at 1 year. Recently, Morales et al.¹⁶⁾ reported that 5-year survival of TAPVR with heterotaxy patients was 79%, which was comparable to nonheterotaxy patients. On the other hand, Hancock et al.³⁾ reported that early mortality and overall mortality in the univentricular group were 36% and 51%, respectively. This was significantly worse than 12% of overall mortality in the biventricular group. In Korea, Lee et al.⁷⁾ reported that early mortality of TAPVR in univentricular heart from 1987 to 2002 was 43.6%, similar to other reports. In our institute, overall mortality of TAPVR in heterotaxy patients was 42.9% from 1991 to 1999.¹⁷⁾ This result is similar with early mortality of 41.2% and overall mortality of 58.8% in this report. However, considering the mean age of 17 months and mean body weight of 7.3 kg at time of surgery in our previous study,¹⁷⁾ we know that the age of operation in this report was significantly earlier, an indication that the condition of patients in this report was more serious and surgical technique was improved. Many authors reported that preoperative pulmonary venous obstruction was related to high mortality,³⁾¹⁷⁾ but in this study there was no statistical significance of preoperative obstruction with mortality ($p=0.162$). However, the most important thing is concomitant palliative surgery. The highest mortality was recorded in patients with modified BT shunt (6 of 7; 85.7%) and the next was in patients with PAB (2 of 3; 66.6%). However, in patients who underwent BCPS and the Fontan procedure, there was no early mortality, and 1 late mortality. Therefore, we know that the TAPVR repair with concomitant palliation of the pulmonary artery is high risk. Morales et al.¹⁶⁾ reported that risk factors for death included diagnosis of pulmonary atresia and need for

shunt operation at the time of TAPVR repair. Lee et al.⁷⁾ reported that a poor preoperative hemodynamic condition and prolonged operation time were risk factors for mortality. In our study, age and body weight at time of surgery was significantly associated with mortality. Preoperative arterial oxygen saturation and medical support were not statistically associated with mortality.

Postoperative pulmonary vein stenosis is an important complication of TAPVR repair in univentricular heart. Hancock et al.³⁾ reported that reoperation of postoperative pulmonary vein stenosis in univentricular heart was 18%, which was significantly high compared with 8% in the biventricular group. In our patients, 2 underwent reoperation due to individual pulmonary vein stenosis, not anastomotic stenosis, and 1 of them died. For postoperative pulmonary vein stenosis, many studies and improvements in surgical and suture techniques have brought improvement to anastomotic site stenosis. However, the histologic anomaly of an individual pulmonary vein considerably increases the risk of postoperative individual vein stenosis.¹⁸⁾¹⁹⁾

In conclusion, There is marked improvement in surgical results from isolated TAPVR with improvement of perioperative diagnosis, treatment, and surgical technique. However, surgical correction of TAPVR combined with univentricular heart is still high risk with high mortality. In particular, concomitant palliation of the pulmonary artery, with early age and low weight at time of surgery were significant risk factors.

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