

Long-term results of one-and-a-half ventricle repair in complex cardiac anomalies

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Abstract

Objective: One-and-a-half ventricle repair is a surgical option for complex cardiac anomalies characterized by right-ventricle hypoplasia or dysfunction. The long-term result analyses or large clinical reviews are rare. The aim of this study is to evaluate the long-term functional results of this surgical procedure. **Methods:** The 29 patients, who underwent one-and-a-half ventricle repair from June 1993 to June 2007, at our Institution, were included. The median age was 26 months (range 6 months to 26 years). One-and-a-half-ventricle repair was performed for volume unloading the small right ventricle (group A, $n = 18$), for work unloading in patients with chronic right-ventricle dysfunction (group B, $n = 9$), and with the acute postoperative right-ventricular dysfunction (group C, $n = 2$). The mean Z value of the tricuspid valve in group A was -3.6 ± 0.7 (range -2.6 to -4.8). The median follow-up duration of hospital survivors was 82 months (range 3 months to 16 years). **Results:** There were four early deaths (two in group A and C, respectively) and no late cardiac death. During follow-up, no patient had superior vena cava (SVC) hypertension or chronic atrial arrhythmia. There was one patient with protein-losing enteropathy. Functional status was New York Heart Association Functional Class I in 21 patients and class II in three patients. Arterial oxygen saturation increased significantly after operation, compared with the preoperative saturation (86.6 ± 9.7 – $96.8 \pm 4.0\%$, $p < 0.01$). Two patients in group B needed medications related to the cardiac function. Four patients underwent reoperation. The 10-year freedoms from late reoperation were $80.0 \pm 12.6\%$ in group A and $51.4 \pm 20.4\%$ in group B. **Conclusions:** The patients with one-and-a-half ventricle repair resulted in favorable late survival in this series. During the follow-up period, most surviving patients showed good functional status without common late complications of the Fontan procedure such as, recurrent cyanosis, pulmonary arteriovenous fistulas, chronic arrhythmias, and SVC syndrome. This procedure appears to be a valid alternative to Fontan and biventricular repairs in patients with right-ventricular dysfunction or hypoplasia.

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Keywords: One-and-a-half-ventricle repair; Right-ventricular hypoplasia; Right-ventricular dysfunction

1. Introduction

Most children with congenital cardiac disease are amenable to either single-ventricle palliation or biventricular repair. There are a few patients, who have a good systemic ventricle, but a pulmonary ventricle that may not be capable of supporting the pulmonary circulation because of ventricular size or function [1,2]. Determining the optimal repair strategy for these patients remains a challenge. Treatment strategies include a Fontan-type procedure, the bidirectional Glenn shunt, or a biventricular repair with atrial septal fenestration.

Long-term outcomes of the Fontan procedure have been less than ideal. Late problems of ventricular hypertrophy, dilation, and eventual failure are time-dependent and appear to be inevitable [3]. At the same time, biventricular

repair in patients with borderline pulmonary ventricle is associated with significant early morbidity and mortality [4].

One-and-a-half-ventricle repair (1.5VR) would be logical if it improves the patient's survival and functional status. The long-term result or large clinical reviews regarding 1.5VR are rare. The aim of this study is to review our experience and to evaluate the long-term functional clinical results of this surgical procedure.

2. Material and methods

2.1. Study population

Between 1993 and 2007, 29 patients underwent bidirectional cavopulmonary shunt (BCPS) with various intraventricular repairs (1.5VR) at Seoul National University Children's Hospital. There were 17 males and 12 females with a median age of 26 months (range 6 months to 26 years).

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Table 1. Underlying primary diagnosis; number of patients and group.

Diagnosis	A	B	C	Total
Ebstein's anomaly	3	4		7
Pulmonary atresia with intact ventricular septum	4		1	5
Atrioventricular septal defect	3	1		4
D-transposition of the great artery	4			4
L-transposition of the great artery with PS		2		2
Tetralogy of Fallot		1	1	2
Double-outlet of right ventricle with PS	2			2
Atrial septal defect	1			1
Double-inlet left ventricle		1		1
Ventricular septal defect	1			1

PS, pulmonary stenosis.

Mean body weight was 17.8 ± 16.1 kg. Median follow-up duration was 82 months (range 3 months to 16 years). We divided these patients according to the classification of Van Arsdell and associates [1]. Group A included 18 patients in whom 1.5VR were performed for volume unloading the small right ventricle (RV). The mean Z value of the tricuspid valve in this group was -3.6 ± 0.7 (range -2.6 to -4.8). Group B consisted of nine patients, who had preoperative RV dysfunction. Group C included 2 patients who underwent 1.5VR as a salvage operation for acute postoperative RV dysfunction after attempted biventricular repair. Primary diagnoses are shown in Table 1 and previous palliative operations in Table 2.

2.2. Surgical technique

All patients underwent median sternotomy. A superior vena cava (SVC) to pulmonary artery (PA) anastomosis was done on cardiopulmonary bypass. We have not performed right PA banding in all cases. The azygos or hemiazygos vein was divided in all cases. Adjustable atrial fenestration was carried out in three patients with high postoperative right-atrium pressure. The concomitant cardiac procedures are shown in Table 3.

2.3. Statistical analysis

Data were analyzed with Statistical Package for Social Sciences (SPSS) 17.0 package (SPSS Inc., Chicago, IL, USA). All values of continuous variable were expressed as mean \pm SD. Chi-square test or Fisher's exact test were applied for comparing and evaluating the operation results. Differences were considered statistically significant when a *p* value was

Table 2. Previous palliative operations.

Operation names	A	B	C	Total
PA banding	3	1		4
Arterial switch operation with PA banding	2			2
Arterial switch operation with coarctoplasty	1			1
Modified B–T shunt	1	1		2
Pulmonary valvotomy and modified B–T shunt	1		1	2
Transannular RVOT widening and modified B–T shunt	1			1
Transannular RVOT widening and central shunt	1			1
Pulsatile bidirectional cavopulmonary shunt		1		1

PA, pulmonary artery; B–T, Blalock–Taussing; RVOT, right-ventricular outflow tract.

Table 3. Concomitant cardiac procedures.

	Group A	Group B	Group C
ASD closure	8	4	
VSD closure	7	1	
Tricuspid valvuloplasty	6	4	
PA angioplasty	5	2	
Previous shunt takedown	5		
PA debanding	4		
RVOT widening	2		1
RA reduction plasty	3	5	
Plication of the aRV	3	2	
ASD fenestration	3		
C-ECD correction	2		1
RV-PA conduit interposition	1		
Aortic valvuloplasty	1		
Mitral valvuloplasty	1		
ASO	1		
PDA ligation	1	1	
Cryoablation		5	
Thrombectomy		2	
TV replacement		2	
PV replacement		1	
Pulmonary valvotomy		1	

ASD, atrial septal defect; VSD, ventricular septal defect; PA, pulmonary artery; RVOT, right-ventricular outflow tract; RA, right atrium; aRV, atrialized right ventricle; C-ECD, common endocardial cushion defect; ASO, arterial switch operation; PDA, patent ductus arteriosus; TV, tricuspid valve; PV, pulmonary valve.

less than 0.05. Analysis of time-related survival and reoperation was performed using the Kaplan–Meier method.

3. Results

3.1. Early results

Postoperative oxygen saturation ranged from 86% to 100% (mean $96.8 \pm 4.0\%$) at hospital discharge. The oxygen saturation increased (mean $10.0 \pm 9.5\%$) after operation, compared with the preoperative saturation. Mean postoperative central venous pressure (CVP) was below 20 mm Hg in all patients except one and ranged from 10 to 25 mm Hg (mean 15 ± 4 mm Hg) at intensive care unit discharge. The patient with high CVP had a mild facial edema during the acute postoperative period but improved at hospital discharge.

Operative death was defined as death associated with any cardiac operation. Operative mortality was 13.8% (two in group A, two in group C). One patient with additional RV to PA conduit died of right-heart failure and another patient with Down syndrome died of hepatic failure in group A. Two patients in group C died of biventricular failure, postoperatively. Group C patients had a statistically significant higher incidence of early death as compared with the other two groups ($p < 0.01$). The detailed causes of deaths are listed in Table 4.

The operation-related complications were chylothorax in 3 cases, arrhythmia in 3 cases, diaphragmatic palsy in 2 cases, acute renal failure in 1 case, and seizure in 1 case. Early reoperations were performed on 2 patients due to postoperative bleeding and complete atrioventricular block, on the operation day and the postoperative day 1, respectively.

Table 4. Hospital deaths.

Patient no.	Group	Age (mo)	Weight (kg)	Diagnosis	Preoperative data and state	Procedures performed	Postoperative data	CPB/ACC time (min)	Cause of death
1	A	10	11	Tetralogy of Fallot, complete AVSD (Rastelli type C)	TV Z value –3.2	AVSD repair, RVOT widening, ASD closure, BCPS	CVP 22 mm Hg LAP 12 mm Hg	218/124	RHF on the POD # 26
2	A	28	8.7	Unbalanced AVSD (Rastelli type C)	TV Z value –3.6	AVSD repair, BCPS PA debanding	CVP 21 mm Hg LAP 10 mm Hg	225/142	Hepatic failure on the POD # 39
3	C	6	7.8	Down syndrome Tetralogy of Fallot, acute RHF	Unstable postoperative hemodynamic state, ventilator care state	BCPS	CPB weaning failure; ECMO support	150/0	RHF on the POD # 3
4	C	12	8.9	PA with IVS, severe AI, acute RHF	TV Z value –3.2	B–T shunt division, PA angioplasty, RV-PA conduit interposition, ASD closure, BCPS, aortic valvuloplasty	CVP 15 mm Hg	498/173	RHF on the POD # 4

AVSD, atrioventricular septal defect; TV, tricuspid valve; RVOT, right-ventricular outflow tract; BCPS, bidirectional cavopulmonary shunt; CVP, central venous pressure; LAP, left-atrial pressure; RHF, right-heart failure; POD, postoperative day; CPB, cardiopulmonary bypass; ECMO, extracorporeal membrane oxygenator; PA, pulmonary artery; RV, right ventricle; AI, aortic insufficiency.

Table 5. Late reoperation.

Patient no.	Group	Diagnosis	Previous operation	Reoperation cause	Reoperation name	Interval to reoperation
1	A	DORV, PS	BCPS, Rastelli operation	RV-PA conduit stenosis	Conduit change	96 months
2	A	Ebstein anomaly	BCPS, TVP plication of the aRV,	Moderate TR, atrial fibrillation, huge aRV	TVP, plication of the aRV, maze procedure	98 months
3	B	ccTGA, PS, VSD	BCPS, pulmonary valvotomy	Moderate TR,	TVR	56 months
4	B	PS, PR, TR	BCPS, TVP PVR with tissue valve	Infective endocarditis	Pulmonary valve replacement	14 months

DORV, double outlet right ventricle; PS, pulmonary stenosis; BCPS, bidirectional cavopulmonary shunt; RV, right ventricle; PA, pulmonary artery; TVP, tricuspid valve plasty; aRV, atrialized right ventricle; TR, tricuspid regurgitation; ccTGA, congenital corrected transposition of the great artery; VSD, ventricular septal defect; TVR, tricuspid valve replacement; PR, pulmonary regurgitation; PVR, pulmonary valve replacement.

3.2. Late results

There was one late death. The patient with Ebstein’s anomaly died 7 months after operation of hepatocellular carcinoma. No patient died of cardiac problem.

No patients had SVC syndrome, chronic arrhythmia, and cyanosis. Protein-losing enteropathy developed in one patient with double-inlet left ventricle at 4 years after operation. The patient underwent 1.5VR for RV dysfunction after ventricular septation. He improved by 2 months after medical treatment, and had no recurrence during the 2-year follow-up period.

Two patients in group A, and two in group B underwent late reoperations. Two patients in group A had tricuspid valve (TV) repair and Rastelli conduit change, respectively. Two patients in group B had TV replacement and pulmonary valve replacement, respectively. The detailed causes of reoperations are listed in Table 5. One patient with severe RV dysfunction is currently awaiting a Fontan procedure. The patient underwent 1.5VR for RV dysfunction and severe tricuspid regurgitation (TR) after repair of partial endocardial cushion defect. However, RV dysfunction and TR advanced again at 6 years after 1.5VR. The 10-year freedoms from late reoperation were 80.0 ± 12.6% in group A, and 51.4 ± 20.4% in group B (Fig. 1). Patients with chronic RV dysfunction required more reoperation (*p* = 0.02).

A total of 24 survivors have been followed up for a mean of 85.6 ± 44.6 months (range 3 to 190 months). No patient died

of cardiac problem during the follow-up period. During follow-up, the functional status of these patients was New York Heart Association (NYHA) Functional Class I in 21 patients and Class II in 3 patients. Two patients in NYHA Functional Class II were group B and they are on medications related to the cardiac function.

At follow-up echocardiography, 2 patients in group A, and 4 in group B had more than mild TR. Postoperative cardiopulmonary exercise test was done in 9 of the surviving 24 patients and maximal oxygen consumption was mean 31.9 ± 5.3 ml kg⁻¹ min⁻¹ (range 21.4 to 38.4 ml kg⁻¹ min⁻¹).

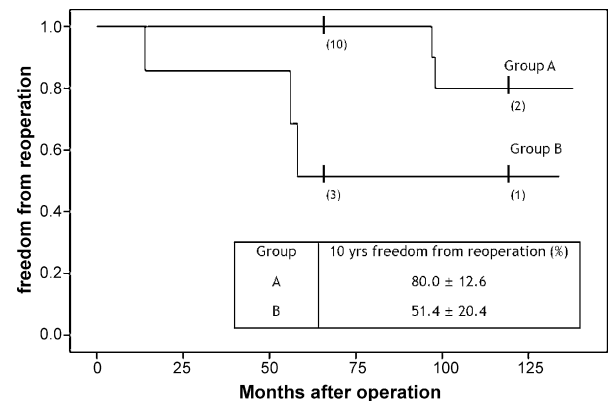


Fig. 1. Freedom from reoperation according to the groups.

4. Discussion

The aim of the 1.5VR is to create a modified in-series circulation, with no intracardiac shunting, the systemic circulation fully supported by the systemic ventricle, and pulmonary circulation dependent on a superior cavopulmonary connection for the SVC return and supported by an insufficient pulmonary ventricle for the inferior vena cava (IVC) return [2,5,6]. This procedure allows the maintenance of equal and separate pulmonary and systemic circulations while, at the same time, incorporating the pulmonary ventricle into the circulation with a reduced volume load [2,5]. There are many advantages in incorporating an insufficient ventricle to partly support the pulmonary circulation, such as ability to increase the cardiac output, adaptation to exercise, maintenance of pulsatile flow in the pulmonary circulation, providing hepatic venous blood to both lungs, flexibility to increased pulmonary vascular resistance, and circulation at low venous pressure in the IVC system [5,6].

This procedure is widely used for patients with a variety of forms of complex congenital heart disease. The disease spectrum includes patients with atrioventricular septal defect, double-outlet right ventricle (DORV), tetralogy of Fallot, D-transposition of the great arteries (TGA), congenital corrected TGA, Ebstein's anomaly, pulmonary atresia with intact ventricular septum, straddling and overriding tricuspid valve, and pulmonary stenosis (PS) [1,5–8]. However, there is controversy for a range of appropriate RV size or function for this approach. Van Arsdell and associates [1] described that patients with a tricuspid Z value between -2 and -10 were considered for this approach. Alvarado and associates [9] also successfully used this approach for tricuspid Z value as small as -10 . Conversely, Chowdhury and associates [10] described that patients with a tricuspid Z value between -1.5 and -4.8 may be suited for this approach. We agree with Muster and associates [11] that not only tricuspid Z value but also right-ventricular compliance, TR, right-ventricular outflow tract, relative PA hypoplasia, and pulmonary vascular resistance are important factors for operative success.

For all patients with borderline RV function or size, the decision to perform 1.5VR need not be made before the operation. Several other authors reported the application of this procedure when there is a borderline-sized RV and an elevated central pressure immediately after bypass following biventricular repair [1,2,5]. In our series, one patient who may be able to tolerate complete biventricular repair underwent Rastelli operation for Fallot-type DORV. However, we added a bidirectional Glenn anastomosis on postoperative 1 day because of persistent low cardiac output and high CVP. The patient was doing well at the 16-year follow-up.

One of the major concerns in the extended application of 1.5VR is the currently limited knowledge of the long-term results, due to the relatively recent use of this surgical approach and to a few clinical reports about this procedure [7]. In this retrospective review of 29 patients undergoing this procedure, we showed that patients with 1.5VR resulted in favorable late outcomes. During the follow-up period, no patient died of cardiac problem and most surviving patients showed good functional status without common late

complications of the Fontan procedure such as, cyanosis, chronic arrhythmias, pulmonary arteriovenous fistulas, and SVC syndrome.

In our initial experience, we used this procedure as a salvage operation for acute postoperative RV dysfunction and the results were unsatisfactory. We agree with previous reports that a cavopulmonary connection as a salvage procedure in treating an acute RV dysfunction during the attempt of a biventricular repair may be usually inappropriate [1,8,10,12]. At present, we use extracorporeal membrane oxygenator instead of this procedure as a salvage operation for acute postoperative RV dysfunction.

We also agree with Chowdhury and associates [8,10] that patients requiring an intricate intracardiac repair with prolonged cardiopulmonary bypass time for dedicating the IVC blood alone to the RV may be considered as having a relative contraindication to 1.5VR in favor of a simpler Fontan-type repair. In our study group, two deaths in group A were associated with prolonged cardiopulmonary bypass time.

Another difficult issue is determination of when to include atrial septal fenestration [8]. Atrial fenestration may be effective in patients with high risks but it carries the problem of chronic cyanosis and paradoxical emboli. In addition, volume loading of the heart continues and the appropriate fenestration size may not be predictable [1,8,13]. Three patients in our study group underwent concomitant atrial septal fenestration due to postoperative high right-atrial pressure (more than 12 mm Hg or twice left-atrial pressure). Postoperative oxygen saturation in these three patients was above 90% at hospital discharge and maximal oxygen consumption at postoperative cardiopulmonary exercise test was mean 33.8 ± 3.2 ml $\text{kg}^{-1} \text{min}^{-1}$. No patients had fenestration-related problems.

There are still many debates on blocking or not blocking the azygos or hemiazygos vein during the 1.5VR. With the bidirectional Glenn shunt, a patent azygos or hemiazygos vein would theoretically provide the advantage of decompressing the SVC or the right atrium in patients with SVC hypertension or severe RV dysfunction. In addition, it would permit blood to flow to the upper compartment during exercise, when the pulmonary ventricle is not capable of handling increased systemic venous return [8,14,15]. However, there have been reports of development of a semicircular circulation in which blood is diverted down and recirculates back to the heart [5,8]. In our study, the azygos or hemiazygos vein was ligated in all cases to avoid systemic venous runoff to the IVC.

In the setting of a 1.5VR, right PA banding proximal to cavopulmonary anastomosis would decrease the excessive pulsatility in the SVC associated with pulmonary regurgitation and to avoid the risk of an aneurysmal SVC [8,14]. However, we have not performed right PA banding in all patients and no patients had SVC aneurysm. Therefore, we think that right PA banding is an unnecessary procedure.

We believe that 1.5VR offers good palliation in selected patients. This procedure has the advantage of producing pulsatile pulmonary blood flow and maintaining a low pressure in the IVC compartment [14]. In the setting of a 1.5VR, the RV has to handle only two-thirds of the venous return, which would reserve RV function for a longer period

[16]. Further follow-up is needed to assess the longer term effects of this procedure on RV function.

There are several limitations to the current study. The ability to generalize the present findings is limited because the study was a retrospective analysis at a single center and was not randomized. Therefore, the results should be interpreted with caution. Randomized, controlled studies are needed to better evaluate the safety and efficacy for 1.5VR.

5. Conclusion

The patients with 1.5VR resulted in favorable late survival in this series. During the follow-up period, most surviving patients showed good functional status without common late complications of the Fontan procedure such as, cyanosis, pulmonary arteriovenous fistulas, chronic arrhythmias, and SVC syndrome. This procedure appears to be a valid alternative to Fontan and biventricular repairs in selected patients with RV dysfunction or hypoplasia.

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