□ 임상연구 □

변형된 노우드(Norwood) 수술의 결과: 단일병원의 지난 7년간의 경험

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Outcome of the Modified Norwood Procedure: 7 Years of Experience from a Single Institution

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Background: We assessed the early and mid-term results of the modified Norwood procedure for first-stage palliation of hypoplastic left heart syndrome (HLHS) and its variants to identify the risk factors for hospital mortality. *Material and Method:* Between March, 2003, and December, 2009, 23 patients (18 males and 5 females) with HLHS or variants underwent the modified Norwood procedure. The age at operation ranged from 3 to 60 days (mean, 11.7±13.2 days) and weight at operation ranged from 2.2 to 4.8 kg (mean, 3.17±0.52 kg). We used a modified technique that spared the anterior wall of the main pulmonary artery in 20 patients. The sources of pulmonary blood flow were RV-PA conduit in 15 patients (group I) and RMBTS in 8 (group II). Follow-up was completed in 19 patients (19/20, 95%) in our hospital (mean 26.0±22.8 months). *Result:* Early death occurred in 3 patients (3/23, 13%), of whom 2 had TAPVC. Fourteen patients underwent subsequent bidirectional cavopulmonary connection (BCPC, stage 2) and seven underwent the Fontan operation (stage 3). Three patients died between stages, 2 before stage 2 and one before stage 3. The estimated 1-year and 5-year survival rates were 78% and 69%, respectively. On multivariate regression analysis, aberrant right subclavian artery (RSCA) and associated total anomalous pulmonary venous connection (TAPVC) were risk factors for hospital mortality after stage 1 Norwood procedure. *Conclusion:* HLHS and its variants can be palliated by the modified Norwood procedure with low operative mortality. Total anomalous pulmonary venous connection adversely affects the survival after a stage 1 Norwood procedure, and interstage mortality rates need to be improved.

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Key words: 1. Congenital heart disease (CHD)
2. Hypoplastic left heart disease
3. Norwood procedure

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INTRODUCTION

Since Norwood, Lang, and Hansen[1] first reported a successful application of palliative staged approach for hypoplastic left heart syndrome (HLHS) in 1983, the outlook for the patients with HLHS has dramatically improved[2,3]. However, the Norwood procedure as a stage I palliation still remains a challenging step with a high mortality[4,5].

The indications of the Norwood procedures have been expanded towards functional single ventricle with arch anomaly and variants of HLHS until recent era by pediatric cardiac surgeons. The Norwood procedure must provide unobstructed systemic blood flow from the right ventricle, relieve obstruction to pulmonary venous return, and limit pulmonary blood flow by a systemic-to-pulmonary shunt. Because the pulmonary circulation is perfused from the systemic circulation, a sudden shift in the resistance ratio between the two vascular beds can cause maldistribution of cardiac output. This phenomenon has been implicated as a major cause of early death in these infants[3].

In the classic Norwood procedure, the main pulmonary artery (MPA) is transected at its proximal portion and the neoaorta is reconstructed with a homograft or other nonviable material, which might limit the growth potential of the neoaorta. In order to solve this problem, some surgeons reconstruct the neoaorta with only autologous tissue[6]. However, besides being technically demanding, this technique can narrow the space for the pulmonary arteries behind the neoaorta and cause left bronchial compression, reducing the success rate of the Fontan procedure. To avoid these problems, we developed technical modification sparing anterior wall of main pulmonary artery in neoaortic reconstruction[7].

The purpose of the present study was to assess the early and mid-term results of the modified Norwood procedure during last 7 years in our hospital.

MATERIAL AND METHOD

1) Patient profiles

Between March 2003 and December 2009, 23 patients (18 males and 5 females) with HLHS or variants underwent the Norwood procedure. Anatomic diagnoses made by echo-

Table 1. Diagnosis and associated cardiac anomalies

Condition	Anatomic diagnosis	No.
HLHS	MA+AA	11
	MA+AA+TAPVC (supracardiac)	1
	MS+AA	2
	MS+AS	2
	MS+AS+IAA	1
	MA+AS+VSD	1
	AA+single ventricle (common AV valve)	1
		19 (82.6%)
Variants	TA (type IIc)+IAA	1
	c-AVSD+IAA+aberrant RSCA +TAPVC (supracardiac)	1
	c-AVSD+CoA	1
	DILV, TGA+CoA	1
		4 (17.4%)

AA=Aortic atresia; AS=Aortic stenosis; c-AVSD=Complete atrioventricular septal defect; AV valve=Atrioventricular valve; DILV=Double inlet left ventricle; HLHS=Hypoplastic left heart syndrome; IAA=Interrupted aortic arch; MA=Mitral atresia; MS=Mitral stenosis; RSCA=Right subcalvian argery; TA=Tricuspid atresia; TAPVC=Total anomalous pulmonary venous connection; TGA=Transposition of great arteries; VSD=Ventricular septal defect.

cardiography and operative findings are listed in Table 1. Nineteen patients had classic HLHS, including aortic atresia or stenosis, mitral atresia or stenosis with a poorly developed left ventricle. The remaining 4 patients had variants of HLHS with left ventricular and aortic arch hypoplasia. The diameters of the ascending aorta ranged from 1.5 to 7.0 mm (mean, 3.17±1.72 mm) and were 2 mm or less in 8 patients (34.8%). Tricuspid regurgitation documented by color flow Doppler echocardiography was mild or more in 12 patients (52.2%). Five patients (21.7%) had aortic arch anomalies (coarctation of aorta or interrupted aortic arch).

Age at operation ranged from 3 to 60 days (mean, 11.7± 13.2 days). Weight at operation ranged from 2.2 to 4.8 kg (mean, 3.17±0.52 kg). Twelve patients (12/23, 52.2%) underwent mechanical ventilation before the operation, and all patients received an infusion of prostaglandin E1.
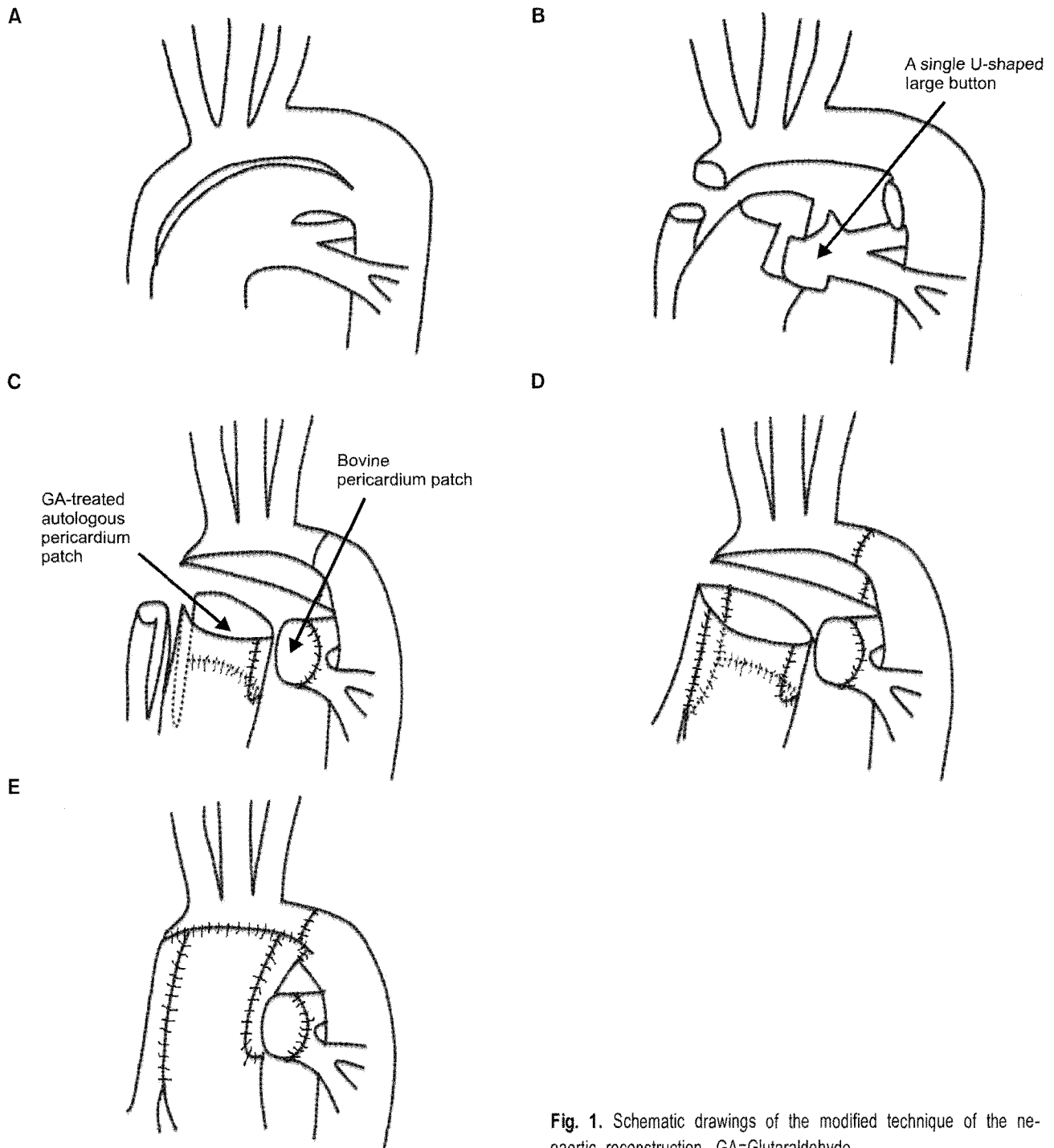
A

B

A single U-shaped
large button

C

GA-treated
autologous
pericardium
patch

Bovine
pericardium patch

D

E

Fig. 1. Schematic drawings of the modified technique of the ne-oaortic reconstruction. GA=Glutaraldehyde.

2) Surgical technique

(1) Cardiopulmonary bypass and technical modification in neoaortic reconstruction: In the first 3 patients, MPA

transection and direct anastomosis to the aortic arch was per-formed during arch reconstruction, and we used our technical modification of the neoaortic reconstruction in other 20 patients. To avoid use of total circulatory arrest, arterial per-

fusion was performed through the innominate artery via a 3- or 3.5-mm polytetrafluoroethylene (PTFE) tube graft for both systemic and regional cerebral perfusions. After bicaval venous cannulation, cardiopulmonary bypass commenced.

We excised both pulmonary arteries from the posterior wall of the MPA in a single U-shaped large button to spare the large anterior MPA wall (Fig. 1B). The two openings of right and left pulmonary arteries were covered with redundant bovine pericardial patch. The pulmonary artery isolation was performed under beating heart. The distal ascending aorta was transected at just proximal to the innominate artery and cardiplegia was then induced by direct infusion of the cold blood cardioplegic solution into the divided small ascending aorta. A long longitudinal incision at the ascending aorta and a small vertical incision at the MPA were made, respectively, at their corresponding sites, and the longitudinally incised ascending aorta was incorporated to the MPA. The defect created after excision of both pulmonary arteries was reconstructed with a small piece of glutaraldehyde-treated autologous pericardial patch. After the neoaortic reconstruction, all head vessels are temporarily occluded and the regional cerebral perfusion commenced at rectal temperature of 25°C (50 mL/kg/min, 30 to 50 mmHg of mean perfusion pressure). The clamp at the PDA was removed, and the aortic isthmus was divided. The aortic arch was completely opened by making a longitudinal incision along the lesser curvature of the aortic arch. The ductal tissue was completely excised from the fully mobilized upper descending aorta. The longitudinally opened aortic arch was then anastomosed to the descending aorta and the reconstructed neoaorta (Fig. 1C~E).

(2) Atrial septectomy and formation of a source of pulmonary blood flow: Atrial septectomy was carried out under fibrillating condition induced by the fibrillator. After the atrial septectomy, a source of pulmonary blood flow was made. Right ventricular right ventricle-pulmonary artery (RV-PA) conduit connection was carried out under fibrillating condition before June 2007 in 14 patients, and the right modified Blalock-Taussig shunt (RMBTS) has been used since then in 9 patients. One patient who had RMBTS required intraoperative conversion to RV-PA conduit due to hypoxemia. Finally, RV-PA conduit was performed in 15 patients (group I), and RMBTS in 8 patients (group II) as a source of pul-

Table 2. Preoperative profiles of each patient group

Variables	Group I (n=15)	Group II (n=8)	p-value
Age (days)	14.1±15.8	7.3±3.0	0.126
Sex (male/female)	11/4	7/1	0.433
Weight (kg)	3.19±0.62	3.14±0.30	0.830
Classic HLHS	12 (80%)	7 (87.5%)	0.651
TR or AVVR (≥mild)	7 (46.7%)	5 (62.5%)	0.469
Ascending aorta diameter (mm)	3.15±1.78	3.21±1.70	0.939
Ascending aorta diameter ≤ 2 mm	6 (40%)	2 (25%)	0.472
Aortic arch anomaly (CoA or interrupted Ao arch)	4 (26.7%)	1 (12.5%)	0.433
Preoperative ventilator care	9 (60%)	3 (37.5%)	0.304
Preoperative PGE1 infusion	All	All	1.000

Ao=Aorta; AVVR=Atrioventricular valve regurgitation; CoA= Coarctation of aorta; HLHS=Hypoplastic left heart syndrome; PGE1=Prostaglandin E1; TR=Tricuspid regurgitation; Group I=RV-PA conduit as a source of pulmonary blood flow; Group II=RMBTS as a source of pulmonary blood flow.

monary blood flow. In the RV-PA conduit group, the size of the shunt used was 5 mm in 5 patients, and 6 mm in 10 patients. In the RMBTS group, all patients was used 3.5 mm PTFE tube graft, and RMBTS was banded using a 3.5 mm PTFE tube graft strip in 5 patients. Preoperative profiles of each patient group are shown in Table 2.

(3) Additional procedures and postoperative management: Five patients underwent concomitant intracardiac repair during the Norwood procedure. These included valvuloplasty of the tricuspid or common atrioventricular valve in 3 patients, and repair of total anomalous pulmonary venous connection (TAPVC) in 2 patients.

The sternum was routinely left open, and the peritoneal dialysis for body water removal was used in all patients. Ventilator settings were adjusted to keep arterial oxygen saturations between 75% and 85% and carbon dioxide levels from 40 to 50 mmHg. Hemoglobin level was kept over 15 g/dL. The inotropic drugs mostly used were dopamine or dobutamine 5 to 10 μg/kg per minute, milrinone 0.35 to 0.7 μg/kg per minute, and calcium gluconate 10 mg/kg per hour. Vasopressin 0.0001 to 0.0006 u/kg per minute was also used to regulate systemic vascular resistance.

Table 3. The early postoperative data of each patient group

Variables	Group I (RV-PA conduit) (n=15)	Group II (RMBTS) (n=8)	Total (n=23)	p-value
CPB (minutes)	230±56	180±27	211±53 (146~327)	0.013
ACC (minutes)	81±25	99±9	87±22 (28~127)	0.133
TCA	1 (35 minutes)	0	1 (35 minutes)	
ASCP (minutes)	53.3±16.9	47.8±7.1	50.9±13.4 (37~83)	0.433
DSC (days)	3.1±1.2	3.3±0.5	3.1±1.0 (1~6)	0.700
Ventilator care (days)	10.3±7.9	10.3±7.1	10.3±7.4 (3.8~28.6)	0.981
LOS (days)	43.0±30.2	27.9±14.9	37.0±25.8 (12~105)	0.208

ACC=Aortic cross clamp; ASCP=Antegrade selective cerebral perfusion; CPB=Cardiopulmonary bypass; DSC=Delayed sterna closure; LOS=Length of hospital stay; RMBTS=Right modified Blalock-Taussig shunt; RV-PA=Right ventricle-pulmonary artery; TCA=Total circulatory arrest.

Table 4. Causes of death after the Norwood procedure

Patient No.	Age at op. (days)	Weight at op. (kg)	Diagnosis	Operation	Cause of death	Postoperative days
1	4	2.20	MS+AS+IAA	Norwood op. with RV-PA conduit	Low cardiac output	6
2	12	3.16	c-AVSD+IAA+aberrant RSCA+TAPVC	Norwood op. with RA-PA conduit +repair of TAPVC	Hypoxemia	4
3	20	2.20	MA+AA+TAPVC	Norwood op. with RV-PA conduit +repair of TAPVC	Lower cardiac output (ECMO weaning failure)	3

AA=Aortic atresia; AS=Aortic stenosis; c-AVSD=Completed atrioventicular septal defect; ECMO=Extracorporeal membrane oxygenation; IAA=Interrupted aortic arch; MA=Mitral atresia; MS=Mitral stenosis; RSCA=Right subclavian artery; TAPVC=Total anomalous pulmonary venous connection.

3) Data collection and statistical analysis

Data were collected and managed using Microsoft Excel 2003 and analyzed using SPSS 11.0 (SPSS, Inc., Chicago, IL). In univariate analysis, categoric variables were compared by using the χ^2 test, and continuous variables by using the Student's t test. For evaluation of risk factors for hospital mortality, variables with a value of p≤0.2 on univariate analysis were entered into a multivariate stepwise logistic regression analysis model. Kaplan-Meier survival curve analysis was used to estimate the late survival rates, and survival curves were compared using the log-rank test. All results are expressed as means±standard deviations, and p-values of less than 0.05 were considered statistically significant. Early mortality was defined as death within 30 days of operation or prior to hospital discharge.

RESULT

1) Early postoperative data

The mean cardiopulmonary bypass (CPB) time was 211±53 minutes (range, 146 to 327 minutes), and the mean aortic cross clamp time was 87±22 minutes (range, 28 to 127 minutes). Antegrade regional cerebral perfusion was used for brain protection in all patients except one in whom total circulatory arrest was used. The mean regional cerebral perfusion time was 50.9±13.4 minutes (range, 37 to 83 minutes). The delayed sternal closure was done 3.1±1.0 days after operation. The mean ventilator care time was 10.3±7.4 days (range, 4 to 29 days), and mean hospital stay was 37.0±25.8 days (range, 12 to 105 days). Early postoperative data of each patient group are shown in Table 3. CPB time was significantly longer in the RV-PA conduit group.

Table 5. Risk factor analysis for hospital mortality

Risk factor	Univariate p-value	Multivariate p-value
Age	0.967	
Sex	0.043	0.060
Weight	0.327	
First 3 cases	0.263	
Preoperative ventilator care	0.590	
Classic HLHS	0.435	
Preoperative TR or AVVR ≥ mild	0.590	
Restrictive ASD	0.328	
Aberrant RSCA	0.008	0.012
Ascending aorta ≤ 2 mm	0.955	
Aortic arch anomaly (CoA or interruption)	0.043	0.060
CPB time	0.114	0.104
ACC time	0.711	
Source of pulmonary blood flow (RV-PA conduit vs. RMBTS)	0.175	0.142
Associated TAPVC	<0.001	<0.001

ACC=Aortic cross clamp; ASD=Atrial septal defect; AVVR= Common atrioventricular valve regurgitation; CPB=Cardiopulmonary bypass; HLHS=Hypoplastic left heart syndrome; RMBTS= Right modified Blalock-Taussig shunt; RSCA=Right subclavian artery; RV-PA=Right ventricle- pulmonary artery; TAPVC=Total anomalous pulmonary venous connection; TR=Tricuspid regurgitation.

Postoperative complications were observed in 5 patients (5/23, 21.7%). Inferior vena cava (IVC) thrombus or right atrium thrombus required additional procedures were observed in 2 patients, prolonged ventilator care more than 3 weeks in 2 patients, and postoperative bleeding required reoperation in one patient.

There were 3 early hospital mortalities (3/23, 13.0%). Among them 2 occurred in the patients with TAPVC. The causes of deaths are depicted in Table 4. We analysed the risk factors for hospital mortality. Univariate analysis identified sex, aberrant right subclavian artery (RSCA), aortic arch anomaly, and associated TAPVC as significant risk factors. On multivariate regression analysis, aberrant RSCA and associated TAPVC were independently associated with hospital mortality after the Norwood procedure (Table 5).

2) Late follow-up data

Late follow-up was achieved in 19 patients (19/20, 95%),

with a mean follow-up duration of 26.0±22.8 months (range, 1 to 66 months). One patient successfully underwent the bidirectional cavopulmonary connection (BCPC) and the Fontan operation at other hospital.

There were 3 late deaths among the 20 discharged patients; 2 late sudden cardiac deaths before the BCPC (6 days and one month after discharges, respectively) and one after BCPC. Late intervention included pulmonary artery balloon angioplasty in 6 patients, balloon angioplasty for recoarctation of aorta in one patient, and RMBTS ballooning in one patient before the BCPC. One patient with aortic atresia with ventricular septal defect completed the Rastelli operation. Fourteen patients underwent the BCPC after a mean interval of 9.0±6.4 months (range, 5 to 30 months). Eleven of the 14 patients who underwent the BCPC required pulmonary artery patch angioplasty. One patient died of subarachnoid hemorrhage (SAH) after sudden cardiac arrest during admission for the cardiac catheterization 14 months after BCPC.

Four patients required additional procedures after the BCPC. Two patients underwent pulmonary artery balloon angioplasty. One patient underwent recoarction repair using subclavian artery flap angioplasty, and one patient had tricuspid valve repair. Seven patients underwent the Fontan operation after the BCPC at mean age 28.8±7.3 months (range, 24 to 43 months). One patient had tricuspid valve repair and one patient underwent pulmonary artery patch angioplasty at the time of the Fontan operation. After the Fontan operation, there was no in hospital and late death. The current status of all patients is shown in Fig. 2. The cumulative survival of the 23 patients undergoing the modified Norwood procedure were 78%, 69%, and 69% at 1, 3, and 5 years after surgery respectively (Fig. 3A). For the patients who underwent a RV-PA conduit during the Norwood procedure (group I), survival at 6 months was 73% and 73% at 1 year compared with 86% at 6 months and 86% at 1 year for those who underwent a RMBTS (group II) (p=0.405, log rank test) (Fig. 3B).

DISCUSSION

In our 7-year experience with our modified Norwood procedure, the early survival rate was 87% (20/23). The 1-year

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                    ┌─────────────────┐
                    │ Norwood operation│
                    │       23        │
                    └────────┬────────┘
              ┌──────────────┴──────────────┐
        ┌───────────┐              ┌──────────────────┐
        │ Survivors │              │ In-hospital death│
        │ 20 (87.0%)│              │    3 (13.0%)     │
        └─────┬─────┘              └──────────────────┘
   ┌──────┬───┴──────┬──────────────┐
┌──────┐┌──────────┐┌────────────────┐┌──────────────┐
│ BCPC ││Waiting   ││Biventricular   ││Interstage    │
│ 14*  ││ BCPC     ││repair (rastelli││death         │
│      ││   3      ││operation)  1   ││   2          │
└──┬───┘└──────────┘└────────────────┘└──────────────┘
   ├──────────┐
   │    ┌───────────────┐
   │    │Fontan operation│
   │    │      7*        │
   │    └───────────────┘
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   │    │ Waiting Fontan │
   │    │      6         │
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        │Interstage death│
        │      1         │
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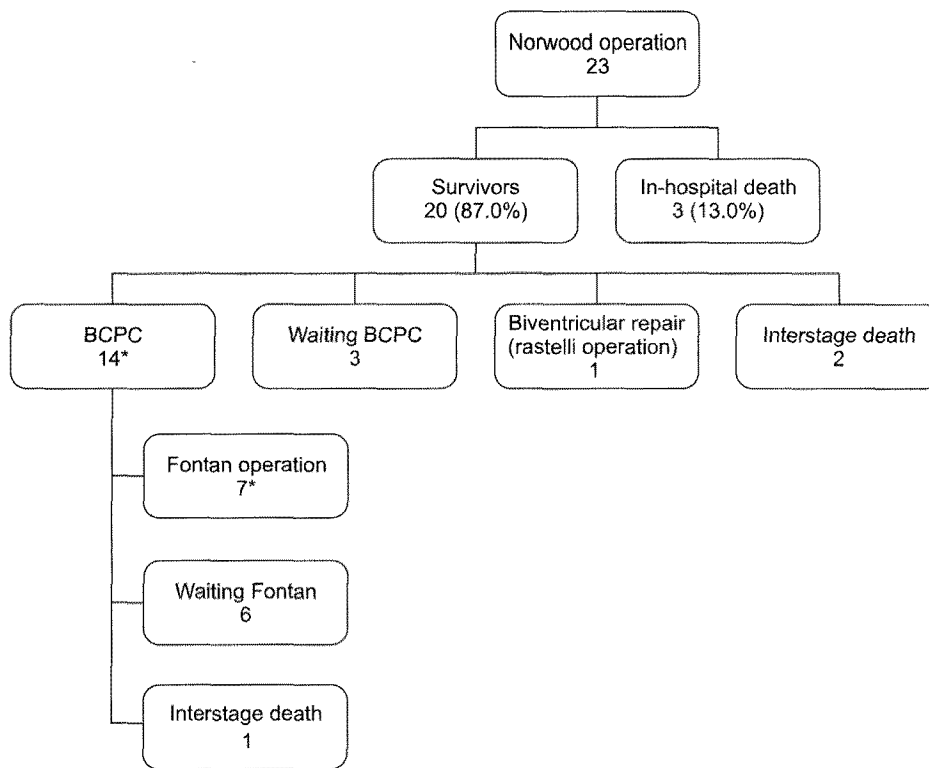
Fig. 2. The current status of all patients after the Norwood procedure for hypoplastic left heart syndrome. BCPC=Bidirectional cavopulmonary connection; *=One patient who underwent the Fontan operation had been transferred to other hospital after the Norwood procedure.

and 5-year survival rates were 78% and 69%, respectively. We found that aberrant RSCA and associated TAPVC were independent risk factors associated with early mortality.

The Norwood procedure has been known as one of the most challenging cardiac anomalies for neonatal cardiac operations, because operative success needs precise anesthetic and cardiopulmonary bypass for brain and myocardial protections and meticulous postoperative management for single ventricular physiology. Only a few centers in North America were able to reproduce some outcomes in the past, but there was a significant improvement in the outcome with the Norwood procedure in recent years. Early mortality was about 30~35% in series in the early to middle 1990s but has improved, and some large centers are now reporting early survival of 85~90%[8-10]. The operative survival of our series showed also good early results (20/23, 87%).

1) The technical modification of the neoaortic reconstruction

Neoaortic reconstruction is an important part in the stage I Norwood procedure. The neoaortic reconstruction using non-viable material can yield several problems, such as no growth potential, calcification, and aneurysmal change[11,12]. The reconstruction without additional patch material can make the short and big neoaorta which can cause pulmonary artery and bronchial compression. Our technique was developed to minimize these future potential problems by incorporating the abundant anterior wall of the MPA to the neoaortic reconstruction. The right and left pulmonary arteries arise together from the posterior wall of the MPA without any gap between the openings of both pulmonary arteries (Fig. 1A). We utilized these anatomic characteristics. By separating both pulmonary arteries from the posterior wall of MPA in the U-shaped button instead of MPA transection we can use its anterior wall in the reconstruction of neoarota. Our technical modifications maximize the use of autologous tissues for the neoaortic reconstruction and save more space for pulmonary arteries and left bronchus behind the neoaorta, which is very important for later successful BCPC and Fontan operation.

2) Factors associated with outcomes

There has been extensive analysis of the risk factors asso-
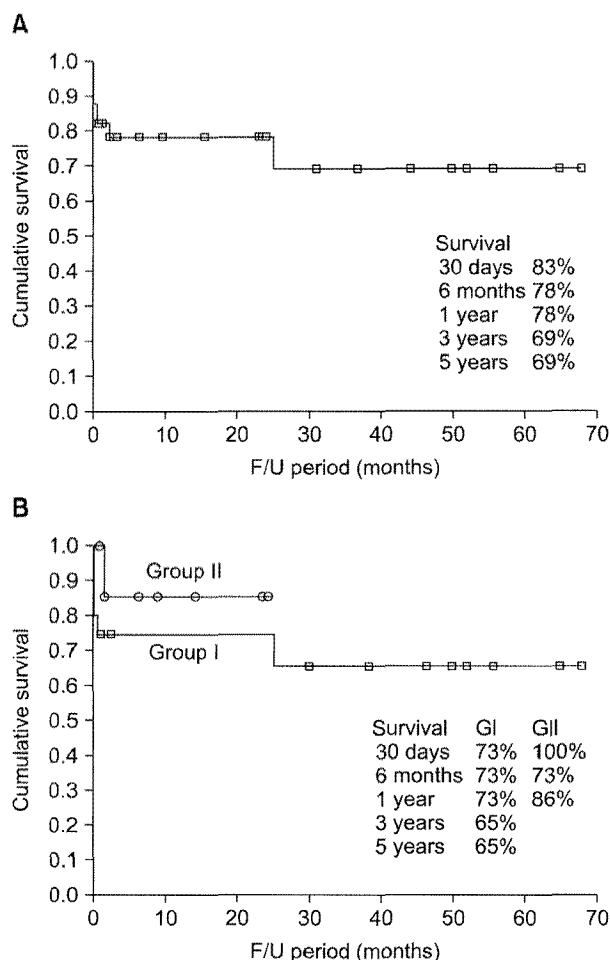
A

B

Fig. 3. Cumulative survival of all patients (A), and each group (B) (p=0.405, log rank test).

ciated with the Norwood procedure. The most frequently cited risk factors are the small size of the ascending aorta[13], low birth weight[14], significant tricuspid regurgitation[15], intact or restrictive atrial septum, and pre-existing impaired ventricular function. The presence of syndrome or other genetic defects is also strongly associated with poor outcome [16]. The anatomic subtypes of HLHS, especially aortic atresia, have been considered as a risk factor for poor outcome[17].

In the present study, we were able to find that aberrant RSCA and associated TAPVC were independent risk factors associated with early mortality. The aberrant RSCA from the descending aorta prevents an extensive mobilization of the descending aorta and thus makes the anastomosis between the MPA and aortic arch difficult[6]. Children with the aberrant

RSCA probably benefit from RV-PA conduit because the patient who has the aberrant RSCA usually has small innominate artery[18]. Hospital survival after the Norwood procedure is significantly worse among the patients with severe obstruction to pulmonary venous return, as reported by Bove and colleagues[19], and the patients with associated TAPVC have more probability of obstruction to pulmonary venous return. Two patients underwent concomitant TAPVC repairs during the Norwood procedure, and all of these patients died 6 days and 3 days after the operation, respectively. One patient died of hypoxemia due to pulmonary hypertension, and the other patient due to low cardiac output and pump weaning failure.

3) Interstage mortality

The persistent incidence of interstage death is of great concern, especially during the first year of life among patients who survived the initial hospitalization. Gaynor and colleagues[20] reported that 12% of the survivors of the initial procedure died before 1 year of age, mostly before the BCPC. These deaths are usually sudden and often unexpected[21]. Residual aortic arch obstruction, restrictive atrial septal defects, imbalance of pulmonary and systemic blood flow, diastolic run-off with coronary ischemia, shunt stenosis of thrombosis, and chronic volume overload of the single ventricle have all been implicated as possible causes for interstage mortality[22]. However, the only identifiable preoperative findings predictive of interstage mortality are restrictive atrial septum and late initial presentation[23].

In this study, the interstage mortality is still significant; 3 (3/20, 15.0%) of the early survivors died before the Fontan completion. The most successful interventions have been the use of home monitoring of pulse oximetry, education of parents to seek advice if oxygen saturations fall below 70%, which might help to pick up early signs of respiratory illness or the insidious development of shunt stenosis or thrombus. This simple measure reduces interstage mortality in one study[24].

4) RV-PA conduit versus RMBTS as a source of pulmonary blood flow

After June 2007, all but one underwent RMBTS as a

source of pulmonary blood flow in the Norwood procedure because we were concerning about the impact of right ventricular incision on late outcome. However, we could not find any difference between RV-PA conduit patients (Group I) and RMBTS patients (Group II) except CPB time. The study to compare RV-PA conduit and RMBTS for the Norwood procedure is ongoing in United States. In the report from Philadelphia by Ballweg and coworkers[25], there is no difference in survival at 3 years. Given that overall survival was not different at the current report, we need more long-term follow-up data from the randomized study to understand which surgical strategy will be better, or more suitable for which subset of patients.

5) Study limitation

Several limitations of the present study require consideration. First, this study involved a relatively small number of patients, which may explain that it is difficult to find some risk factors for mortality, but we were able to find early and mid-term results themselves for our modified Norwood procedure. Second, follow-up periods were different between RV-PA conduit patients and RMBTS patients (RV-PA conduit; 38±22 months versus RMBTS; 10±9 months, p=0.002). The RV-PA conduit group included the early experiences in our institution. However, we were able to compare the early results between RV-PA conduit patients (Group I) and RMBTS patients (Group II). Third, the effect of general improvement in surgical technique and perioperative care as experience accumulated could not be analyzed in the study.

CONCLUSION

In conclusion, the modified Norwood procedure can be performed for the first stage palliation of the hypoplastic left heart syndrome and its variants with low operative mortality comparable to the outcomes from other large centers. Our technical modification sparing the anterior wall of MPA is feasible in the neoaortic reconstruction of the Norwood procedure. An association of total anomalous pulmonary venous connection adversely affects the outcome of the Norwood procedure. More efforts should be paid to reduce the interstage mortality.

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=국문 초록=

배경: 본 연구의 목적은 지난 7년 동안의 좌심형성부전증후군 및 그 변종에 대한 변형 노우드(Nor-wood) 수술의 조기 및 중기 결과를 알아보고, 조기 병원 사망에 영향을 주는 인자들을 확인하는 것이다. 대상 및 방법: 2003년 5월부터 2009년 12월까지 좌심형성부전증후군 및 그 변종으로 노우드 수술을 시행 받은 23명의 환자들을 대상으로 하였다. 수술 당시 나이는 3일에서 60일이었고, 체중은 2.2 kg에서 4.8 kg의 소견을 보였다. 주폐동맥의 전벽을 보존하는 변형된 술식을 20명의 환자에 있어서 적용하였다. 폐동맥혈류 유지는 15명에서 우심실-폐동맥 도관술, 8명에서 우측변형 브라록-타우시히 단락을 이용하였다. 19명(19/20, 95%)의 환자들에 대하여 평균 26.0±22.8개월 동안 추적관찰을 시행 하였다. 결과: 3명(3/23, 13%)의 조기 병원 사망이 있었다. 이중 2명은 폐정맥연결이상을 동반하고 있었다. 14명의 환자가 양방향성 상대정맥 폐동맥 단락술을 시행 받았고, 7명의 환자가 Fontan 수술을 시행 받았다. 양방향성 상대정맥 폐동맥 단락술 이전에 2명의 사망이 있었고, Fontan 수술이전에 1명의 사망이 있었다. 1년과 5년 장기 생존률은 각각 78%와 69%의 소견을 나타냈다. 이상 우측 쇄골하 동맥과 동반된 전 폐정맥연결이상이 병원 사망과 관련된 위험인자들로 나타났다. 결론: 좌심형성부전증후군 및 그 변종에 대한 변형 노우드(Norwood) 수술은 낮은 수술 사망률로 시행될 수 있었다. 전 폐정맥연결이상의 동반이 노우드 수술 후의 생존에 주요한 위험인자임을 알 수 있었다. 향후 중간단계 사망에 대한 성적향상의 노력이 필요할 것으로 판단된다.

중심 단어 : 1. 선천성심장병
　　　　　 2. 좌심형성부전증후군
　　　　　 3. 노우드 수술