

## Outcome of Staged Repair of Tetralogy of Fallot with Pulmonary Atresia and a Ductus-dependent Pulmonary Circulation: Should Primary Repair Be Considered?

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**Background:** The tetralogy of Fallot (TOF) with pulmonary atresia (PA) and a ductus-dependent pulmonary circulation (no major aorto-pulmonary collateral arteries (MAPCAs)) has been treated with staged repair or primary repair depending on the preference of surgeons or institutions. We evaluated the 19-year outcome of staged repair for this anomaly to find out whether our surgical strategy should be changed. **Materials and Methods:** Forty-four patients with TOF/PA with patent ductus arteriosus (PDA) who underwent staged repair from June 1991 to October 2010 were included in this retrospective study. The patients with MAPCAs were excluded. The average age at the first palliative shunt surgery was  $40.8 \pm 67.5$  days (range: 0~332 days). Thirty-one patients (31/44, 70%) were neonates. The average weight was  $3.5 \pm 1.6$  kg (range: 1.6~8.7 kg). A modified Blalock-Taussig (BT) shunt was performed in 38 patients, classic BT shunt in 4 patients, and central shunt in 2 patients. Six patients required concomitant procedures: pulmonary artery angioplasty was performed in 4 patients, pulmonary artery reconstruction in one patient, and re-implantation of the left pulmonary artery to the main pulmonary artery in one patient. Four patients required a second shunt operation before the definitive repair was performed. Thirty-three patients underwent definitive repair at  $24.2 \pm 13.3$  months (range: 7.3~68 months) after the first palliative operation. The average age at the time of definitive repair was  $25.4 \pm 13.5$  months (range: 7.6~68.6 months) and their average weight was  $11.0 \pm 2.1$  kg. For definitive repair, 3 types of right ventricular outflow procedures were used: extra-cardiac conduit was performed in 30 patients, trans-annular patch in 2 patients, and REV operation in 1 patient. One patient was lost to follow-up after hospital discharge. The mean follow-up duration for the rest of the patients was  $72 \pm 37$  months (range: 4~160 months). **Results:** Ten patients (10/44, 22.7%) died before the definitive repair was performed. Four of them died during hospitalization after the shunt operation. Six deaths were thought to be shunt-related. The average time of shunt-related deaths after shunt procedures was 8.7 months (range: 2 days~25.3 months). There was no operative mortality after the definitive repair, but one patient died from dilated cardiomyopathy caused by myocarditis 8 years and 3 months after the definitive repair. Five-year and 10-year survival rates after the first palliative operation were 76.8% and 69.1%, respectively. **Conclusion:** There was a high overall mortality rate in staged repair for the patients with TOF/PA with PDA. Majority of deaths occurred before the definitive repair was performed. Therefore, primary repair or early second stage definitive repair should be considered to enhance the survival rate for patients with TOF/PA with PDA.

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3. Pulmonary atresia

## INTRODUCTION

The tetralogy of Fallot (TOF) with pulmonary atresia (PA) is one extreme group among the wide morphologic spectrum of TOF. The patients in this group of cardiac anomalies cannot survive without a patent ductus arteriosus (PDA) if they do not have aorto-pulmonary collateral arteries. This extreme group of TOF has been traditionally managed with the shunt procedure in the neonatal period or early infancy followed by a subsequent definitive repair due to relative unavailability or short longevity of a small extracardiac conduit, technically more demanding early primary repair, and difficult post-operative management. However, some centers have started to perform a primary one-stage repair for this group of cardiac anomalies since about 20 years ago [1-4]. Although the number of centers performing a primary one-stage repair is increasing, a large number of centers are still doing staged repair because of technical ease of the operation and relative unavailability of small extracardiac conduit for neonates. However, staged repair has several disadvantages: longer exposure to hypoxemia, abnormal growth of pulmonary alveoli and vascular structures, pulmonary artery distortion, right ventricular hypertrophy or even fibrosis due to pressure and volume load on the right ventricle, and a significant attrition rate due to the shunt problem between stages. The aim of this study is to determine whether our surgical policy for the TOF with PA and ductus-dependent pulmonary circulation should be changed based on the results of staged repair over the last 19 years.

## MATERIALS AND METHODS

A retrospective study was conducted based on the medical records of 44 patients with TOF/PA with PDA treated with staged repair in the Department of Thoracic and Cardiovascular Surgery at Dong-A University Hospital and Pusan National University Hospital between June 1991 and

October 2010. The patients with major aorto-pulmonary collateral arteries (MAPCAs) were excluded. Data of survival and death were obtained from the National Statistical Office. The average age at the first systemic-pulmonary shunt operation was  $40.8 \pm 67.5$  days (range: 0~332 days). Among them, 31 patients (70.5%) were neonates, and 39 patients (88.6%) were younger than 90 days of age. The average weight at the time of the shunt operation was  $3.5 \pm 1.6$  kg (range: 1.6~8.7 kg). Associated significant intracardiac anomalies were left pulmonary artery (LPA) sling and partial anomalous pulmonary venous return (PAPVR) in one patient each. Extra-cardiac anomalies were left lung hypoplasia in 1 patient, esophageal atresia with anal atresia in 1 patient, anal atresia in 1 patient, hypospadias with Hirschsprung's disease in 1 patient, and CATCH22 syndrome in 2 patients. All but one had confluent pulmonary arteries. One patient had absence of intrapericardial pulmonary arteries with bilateral PDA and situs inversus. The main pulmonary artery and pulmonary annulus could be inspected in 32 patients. Among them, thirty patients had a main pulmonary artery and 27 of these 30 patients had no pulmonary valve. Three patients had a membranous PA. Two patients did not have a main pulmonary artery. At first palliation, a right modified Blalock-Taussig (RMBT) shunt was performed in 33 patients, right classic Blalock-Taussig (RCBT) shunt in 3 patients, left modified Blalock-Taussig (LMBT) shunt in 5 patients, left classic Blalock-Taussig (LCBT) shunt in 1 patient, and central shunt in 2 patients. Six patients required concomitant procedures: pulmonary artery angioplasty was performed in 4 patients, pulmonary artery reconstruction in 1 patient, and re-implantation of the left pulmonary artery to the main pulmonary artery in 1 patient. Four patients required another systemic-pulmonary shunt or revision before the definitive repair was performed. Three of them required a second procedure due to hypoplastic LPA: one underwent an LMBT shunt as a second shunt operation at 17.4 months after the first shunt operation, and the other two patients underwent a shunt

**Table 1.** Causes of second shunt procedures

Patient no.	Previous operation	Second shunt procedure	Time after 1st operation (months)	Reasons for 2nd operation
1	RMBTS	LMBTS	17.4	Hypoplastic LPA with juxtaductal stenosis
2	RMBTS with LPA angioplasty	Shunt change with LPA angioplasty	5	Hypoplastic LPA with LPA restenosis
3	RMBTS with LPA angioplasty	Shunt change with LPA angioplasty	8	Hypoplastic LPA with LPA restenosis
4	PA conduit reconstruction with central shunt	PA conduit change with LMBTS	6	PA conduit stenosis

RMBTS=Right modified Blalock-Taussig shunt; LMBTS=Left modified Blalock-Taussig shunt; LPA=Left pulmonary artery; PA=Pulmonary artery.

**Table 2.** Types of conduits used in definitive operations in our hospital (n=25/27 patients)

Type of conduit	No. of patient
Polystan porcine pericardium valved conduit	10
Selhigh porcine valved conduit	4
Homemade gore-tex valved conduit	11
Monocusp	3
Bicusp	7
Tricusp	1
Total	25

Six patients (6/33) were excluded because they underwent definitive repair at the other centers.

revision (change) with LPA angioplasty at 5 months and 8 months after the first shunt operation respectively due to re-stenosis of the concomitant LPA angioplasty site of the first RMBT shunt procedure. The remaining one patient was the case with absence of intrapericardial pulmonary artery. A PA conduit change with LMBT shunt was performed at 6 months after the first operation (Table 1).

A total of 33 (75%) of 44 patients underwent definitive repair. Six of those 33 patients underwent definitive repair at other centers. The average age at the time of definitive repair was 25.4±13.5 months (range: 7.6~68.6 months) and the average weight was 11.0±2.1 kg (n=27). The time interval between definitive repair and the first shunt operation was 24.2±13.3 months (range: 7.3~68 months). Right ventricular outflow tract (RVOT) reconstruction with extra-cardiac conduits for the definitive repair was performed in 30 patients, transannular patch repair in 2 patients, and an REV-type operation in 1 patient. Ventricular septal defect (VSD) patch

**Table 3.** Conduit changes after the definitive repair (n=4)

Patient no.	Period after the definitive repair	Operation
1	5 years and 1 month	Conduit change with VSD closure
2	4 years and 6 months	RVOT reconstruction without valve (peel operation)
3	7 years	PVR (peel operation)
4	8 years and 4 months	PVR (peel operation)

VSD=Ventricular septal defect; RVOT=Right ventricular outflow tract; PVR=Pulmonary valve replacement.

fenestration was required in 2 patients. Polystan porcine pericardium valved conduits were used in 10 patients, Selhigh porcine valved conduits in 4 patients, and homemade Gore-Tex valved conduits in 11 patients (Table 2). Various sizes of conduits were used: 14 mm in 2 patients, 16 mm in 11 patients, and 18 mm in 12 patients. Four patients required an RVOT conduit change at 54, 61, 84, and 100 months after the definitive repair (Table 3). Follow-up data was obtained in 43 patients, and the average duration of follow-up was 72±37 months (range: 4~160 months). One case was interpreted as a loss to follow-up. This patient had been discharged from the hospital without any significant problems, but no more information could be collected because the parents of the patient did not cooperate in providing any information after discharge.

All results are expressed as means±standard deviations and the Kaplan-Meier model was used to analyze the survival rate and freedom from extra-cardiac conduit change.

**Table 4.** Causes of hospital deaths

Operation date	Age	Weight (kg)	Associated anomaly	Time to death	Causes of death	Shunt-related death
1994-03-22	13 days	2.6	None	2 days	Sepsis, very small PA, hypoxemia	(+)
1995-08-14	14 days	2.6	None	53 days	Lung problem	(-)
1996-03-29	7 days	3.6	Left lung hypoplasia	37 days	Multiorgan failure	(-)
2004-03-22	3 months	2.1	LPA sling with left main tracheal stenosis	2 hours	Sudden cardiac arrest (bradycardia)	(+)

LPA=Left pulmonary artery; PA=Pulmonary artery.

**Table 5.** Inter-stage deaths between the shunt operation and definitive repair

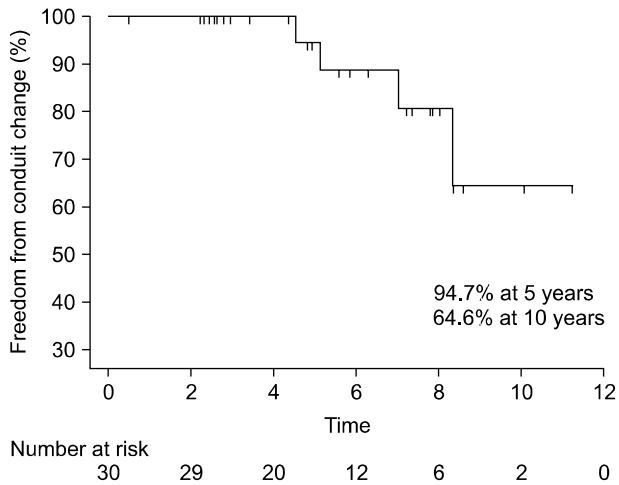
Operation date	Age at operation	Weight (kg)	Associated anomaly	Time to death (months)	Modes of death	Shunt-related death
1991-06-01	13 days	2.2	None	35.5	During admission for cardiac catheterization Brain damage after asphyxia	(-)
1996-01-30	4 months 27 days	7.8	None	16.5	DOA (sudden death)	(+)
1996-08-08	2 months	4.0	None	0.9	DOA (sudden death)	(+)
1997-01-10	9 months	7.8	None	3.9	Nephrotic syndrome, pneumonia	(-)
2005-09-20	4 days	3.03	None	25.3	Sudden death	(+)
2007-12-26	23 days	2.18	Esophageal atresia, anal atresia	7.6	Death during colostomy repair	(+)

DOA=Death on arrival.

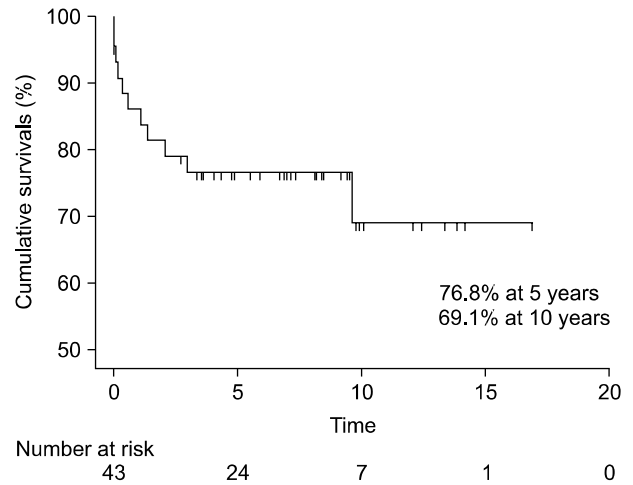
## RESULTS

There were 4 hospital deaths (9.1%) after the shunt procedure. Two were considered shunt-related deaths. One of them underwent a central shunt and reimplantation of the LPA for subcarinal LPA sling with left main bronchus stenosis. She was a premature low birth weight baby (1,190 g at birth, and 2,100 g at operation). Soon after arrival at the intensive care unit (ICU) after surgery, bradycardia and hypotension developed and in the end she died despite cardio-pulmonary resuscitation (CPR). Another one had a severely hypoplastic pulmonary artery. Even after the shunt operation, mild hypoxemia persisted, and as a consequence, the patient died of hypoxemia 2 days after surgery. There were 2 non-shunt-related hospital deaths. One died of lung problems during long-term ventilator care at 53 days after operation. The other patient had a hypoplastic left lung and had been given ventilator care since birth, and was never weaned from it. The death was caused by multiple organ failure followed

by sepsis at 37 days after operation (Table 4). Six patients (6/43, 14.0%) died between the shunt operation and definitive repair, known as inter-stage death. Four deaths were considered to be shunt-related deaths. The inter-stage deaths occurred at 0.9, 7.6, 16.5, and 25.3 months after the shunt operation. Three of them were sudden deaths. One patient died soon after colostomy repair. Two cases were not related to shunt operations. One patient died of hypoxic brain damage caused by asphyxia during hospitalization for cardiac catheterization at 35.5 months after shunt operation. Another patient died of nephrotic syndrome and pneumonia at 3.9 months after shunt operation (Table 5). Six deaths (13.6%) among the overall 10 mortalities (22.7%) were considered to be shunt-related. Shunt-related deaths occurred at an average period of 8.7 months (range: 2 days~25.3 months) after the shunt operation. Definitive repair was performed in 33 (75%) out of a total of 44 patients at an average period of 24.2 months (range: 7.3~68 months) after the shunt operation, and there was no operative mortality after the definitive



**Fig. 1.** Freedom from conduit change.



**Fig. 2.** Cumulative survivals.

repair. One patient died from dilated cardiomyopathy caused by myocarditis at 8 years and 3 months after the definitive repair. In the 25 patients who had undergone the extra-cardiac conduit technique at our center, freedom from extra-cardiac conduit change was 94.7% at 5 years and 64.6% at 10 years (Fig. 1). Overall, the five-year survival rate was 76.8% and 10-year survival rate was 69.1% (Fig. 2).

## DISCUSSION

Staged surgical management for the spectrum of TOF has been a standard procedure for many years. Particularly, the staged surgical approach has been recommended for infants under 1 year of age and infants with unfavorable anatomy [5,6]. However, the debate over staged repair and early total correction continues. Therefore, surgical data of 44 patients with TOF/PA with PDA who received staged surgical management in the last 19 years were studied to clarify the best surgical strategy for this specific cardiac anomaly.

Since Blalock and Taussig introduced the palliative BT shunt, this surgical management technique has been actively performed and has saved the lives of a large number of patients with complex heart diseases and decreased pulmonary blood flow. It has been considered to have several advantages such as low risk, effective palliation, and pulmonary artery growth [7]. In 1981, it was reported that the BT shunt is preferred because it is safe, gives good sustained clinical results,

and has low mortality and good growth of the pulmonary arteries without an increase in pulmonary vascular resistance [8]. After the modified BT shunt was developed, the palliative shunt operation has been accepted as a standard procedure for this congenital heart disease. For about half a century, the modified BT shunt has been the most effective palliative procedure due to its advantages, including a technically simpler method with less dissection and better blood flow to the respective arm compared to the classic BT shunt [9]. In this study, the modified BT shunt was performed as the first stage palliation in most patients (38/44, 86.4%). We adopted the classic BT shunt in small neonates and patients with a very small pulmonary artery during the early phase of our series.

However, as time has passed, the risk associated with BT shunt has been studied and serious problems due to the shunt operation have been discussed. After the shunt operation, for whatever reason, management of decreased pulmonary blood flow is technically demanding [10]. The major cause of decreased pulmonary blood flow is known to be shunt obstruction. Shunt thrombosis and occlusion could give rise to sudden and fatal conditions, and increased-morbidity and mortality after the modified BT shunt are associated with factors such as age, pulmonary artery diameter, and baseline cardiac anatomy [11]. Shunt failure followed by shunt occlusion is infrequent but once it occurs, it often leads to a life-threatening condition [12]. This problem is far more significant in

TOF with pulmonary atresia unlike TOF with PS, which has an alternative pulmonary blood source. In our study, the major causes of death before the definitive repair were shunt-related. More specific causes of hospital deaths are described in Table 4. This shows that half of hospital deaths (2/4) seemed to be shunt-related. After the initial palliative shunt operation, 6 patients (6/43, 14.0%) died before the definitive repairs was performed, which has been called inter-stage death or attrition. Four of the 6 cases of inter-stage death were sudden deaths. Shunt failure due to shunt occlusion is strongly suspected in these cases. Therefore, it can be said that the status after the shunt operation is not safe, and it can significantly contribute to the overall morality in patients with TOF/PA with PDA.

Due to the problems mentioned above, although the BT shunt and modified BT shunt could be considered a low-risk management option for palliation for the spectrum of TOF, it can pose a significant challenge in patient care [13]. As a consequence, early one-stage total repair or early definitive repair after the initial palliative surgery is being discussed as an alternative to staged repair.

Advantages of early definitive repair include removal of the stimulus for right ventricular hypertrophy, alleviation of cyanosis and preservation of myocardial mechanical and electrical function. Early definitive repair is also helpful to avoid the risks associated with shunt operation such as an unstable hemodynamic status due to excessive or insufficient pulmonary blood flow in the immediate postoperative period, sudden inter-stage mortality due to occlusion or critical stenosis of the shunt, distortion of the pulmonary artery branch at the shunt insertion site [2,11,14]. Recently, excellent outcomes of one-stage total repair have been reported. In 1999, a clear survival advantage of primary repair was demonstrated by Van Arsdell and Yun [14]. In the same year, Pigula and colleagues reported that early complete repair could be accomplished with low mortality and low freedom from reoperation in neonates [15]. Moreover, the problems of challenging postoperative care and limitation of conduit size used are also getting solved by the evolution of surgical and medical management, such that the morbidity and mortality of early complete repair for the spectrum of TOF has improved [4]. Furthermore, early complete repair also has short-term bene-

fits (low risk for reoperation and re-intervention), excellent intermediate survival, and long-term benefits (better right ventricular function and lower arrhythmia) [1,4,15]. A comparative study between staged repair and the early complete repair was reported by Park and colleagues [4]. However, early complete repair for small neonates or neonates with a small pulmonary artery is still challenging because of the difficult operative techniques and shortage of adequate extracardiac conduits. We believe that the RV-PA conduit, instead of complete repair, can be an alternative for short-term palliation [16].

Another important conclusion could be derived from Table 5, the column for 'time to attrition'. A total of four shunt-related attritions (inter-stage deaths) were reported and their times to attrition were 16.5, 0.9, 25.3, and 7.6 months. In other words, if the second-stage definitive repair was conducted earlier than 6 months after the first-stage palliative shunt operation, 3 patients could have survived.

In conclusion, there are two surgical strategies that are recommended in patients with TOF/PA with PDA to increase the overall survival rate. Firstly, an early complete repair could be a favorable option. Secondly, in staged management, earlier second-stage definitive repair after the palliative shunt procedure should be considered, or another palliative procedure other than the shunt procedure, such as an RV-PA conduit, should be adopted.

### 1) Study Limitations

Several limitations of the present study require consideration. In our study, we did not compare the outcome of primary repair and the outcome of staged repair. Therefore, we do not vouch for the benefit of early primary repair over staged repair, but we did note that mortality was high after the first palliative operations. Several authors have reported the benefits of early complete repair for the spectrum of TOF [1,4,14,15]. We concluded that primary repair for TOF/PA and ductus dependent pulmonary circulation is a good option. The other limitation is that our study does not include patients with diminutive pulmonary arteries.

## CONCLUSION

The overall survival rate in the patients with TOF/PA with PDA was relatively low after staged repair due to high inter-stage mortality after the shunt operation. Therefore, early complete repair or early second-stage definitive repair after the palliative shunt procedure should be considered to increase the overall survival rate in patients with TOF/PA with PDA.

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