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[UCI]I804:48009-200000175550

Master of Cardiovascular and Thoracic Surgery

Surgical Strategies for Neonates and Young Infants with Pulmonary Atresia and Ventricular Septal Defect: Staged Repair vs. Primary Repair.

신생아 및 조기 영아에서의 심실중격결손을 동반한 폐동맥 폐쇄에 대한 외과적 전략: 단계별 교정술과 일차 완전 교정술에 대한 비교

The Graduate School of the University of Ulsan

Department of Cardiovascular and Thoracic Surgery

Seung Ri Kang

Surgical Strategies for Neonates and Young Infants with Pulmonary Atresia and Ventricular Septal Defect: Staged Repair vs. Primary Repair.

Supervisor: Young Hwue Kim

A Master's Thesis

Submitted to the Graduate School of the University of Ulsan
In Partial Fulfillment of the Requirements
for the Degree of

Master of Cardiovascular and Thoracic Surgery
By

Seung Ri Kang

Department of Cardiovascular and Thoracic Surgery
Ulsan, Korea
December 2018

Surgical Strategies for Neonates and Young Infants with Pulmonary Atresia and Ventricular Septal Defect: Staged Repair vs. Primary Repair.

This certifies that the master's thesis of Seung Ri Kang is approved

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Committee Chairman Professor Young Hwue Kim

Committee Member Professor Tae-Jin Yun

Committee Member Professor Chun Soo Park

Department of Cardiovascular and Thoracic Surgery

Ulsan, Korea

February 2019

Abstract

Background: Initial surgical options for pulmonary atresia with ventricular septal defect (PA/VSD) in neonates and young infants are either palliative shunt operation or early total correction. Staged repair strategy may be associated with a higher risk of inter-stage mortality, and primary repair strategy may lead to frequent post-repair reinterventions.

Methods: From 2004 to 2017, 65 patients with PA/VSD who underwent surgical intervention before the age of 90 days were identified and enrolled in this retrospective study. The cohort was divided into two groups; group R who underwent primary repair (n=15), and group P who underwent Blalock-Taussig shunt or right ventricle to pulmonary artery (RV-PA) conduit (n=50). Risk of multiple reoperations among the survivors was analyzed using Prentice-Williams-Peterson (PWP) model, and Cox proportional hazards model was fitted to determine the risk factors for decreased time to composite adverse outcome (death, reoperations) after birth.

Results: Median follow-up was 42.9 (IQR 19.4-98.7) months. Two groups did not differ in age at initial operation (Group R: 36.5 ± 27.9 days, Group P: 28.4 ± 15.6 p=0.298), BSA

(Group R: 0.2 ± 0.0, Group P: 0.2±0.0, p=0.105), but differed in pulmonary artery index (Group R: 164.5±51.9 mm2/m2, Group P: 124.6±50.9 mm2/m2, p=0.010). During the follow-up duration, there were nine surgical mortalities (Group P=7, Group R=2), 24 first reoperations after repair (Group P=16, Group R=8), and 11 second reoperations (Group P=3, Group R=8). Five-year survival was comparable between the two groups (group R=86.7%, group P=83.6%, p=0.754), but staged repair showed a decreased risk of multiple reoperations compared to primary repair (HR 2.73, 95% CI:0.2751-0.7204, p=0.0086). Cox model showed primary repair as the only risk factor for decreased time to death/first reoperations (HR 2.53, 95% CI:1.002-5.453; P=0.049) and death/second reoperations (HR 2.90, 95% CI:1.09-7.75, p=0.026) after birth.

Conclusions: Staged repair strategy, as compared to initial total repair, was associated with higher inter-stage mortality with less frequent reinterventions after repair, which may be attributable to the use of larger conduits upon repair. Lowering the inter-stage mortality in the staged repair may allow for better surgical outcome in the future.

Keywords: Tetralogy of Fallot, Pulmonary Atresia, Shunt operation,

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Introduction

Pulmonary atresia with ventricular septal defect (PAVSD), is a congenital heart defect (CHD) characterized by discontinuity from the right ventricle (RV) to the pulmonary arteries (PA) as well as a malaligned ventricular septal defect (VSD) caused by anterior displacement of the conal septum. [1] With the exception of PAVSD patients with multiple aortopulmonary collateral arteries (MAPCA), simple forms of PAVSD have ductal dependent pulmonary circulation, requiring early surgical intervention, options of which have been debated. Although all options, namely total correction and palliative shunt, are aimed at restoring sufficient pulmonary circulation, both options entail considerations that must be considered. Early total correction may prevent progression of right ventricular hypertrophy (RVH), while it presents risks of cardiopulmonary bypass (CPB) in the neonatal period, possible diastolic dysfunction of the RV in the presence of small PA's, and the inevitable placement of a small RV-PA conduit, which can lead to a shortened period to a routine conduit change. A palliative systemic-pulmonary shunt, on the other hand, requires concern regarding the immediate postoperative stages in the intensive care unit (ICU), intermediate stage

mortality/morbidity, and the need for additional total repair.

In the current series, with the interest of evaluating the two different surgical strategies, we analyzed neonatal PAVSD patients who underwent surgery at our institution. We sought to analyze the surgical outcomes, long-term rate of reoperation/reintervention, pulmonary artery growth, and the possible risk factors related to such between the 2 surgical strategies.

Methods

Patients

From 2004 to 2017, 65 patients with PA/VSD who underwent surgical intervention before the age of 90 days were identified and enrolled in this retrospective study. All patients had a ductus-dependent pulmonary circulation before their initial operation, and all patients with MAPCAs were excluded. The cohort was then divided into two groups; group R who underwent primary repair (n=15), and group P who underwent Blalock-Taussig (BT) shunt or right ventricle to pulmonary artery (RV-PA) conduit (n=49).

Follow-up

Complete follow-up was complete in 92.3% (60/65) of patients. Mean follow-up duration was 66.7 months. We reviewed all echocardiographic as well as computed tomography (CT) data conducted at our institution for the analysis of pulmonary artery size in order to calculate the Nakata index.

Operations

As for group P (n=49), the most common form of palliation was the right modified BT shunt

(RMBT, n=42, followed by RV-PA conduit (n=3), left modified BT shunt (LMBT, n=2), and central shunt (n=2). All BT shunts utilized a thoracotomy approach via the 4th intercostal space (ICS), and all central shunts via a full median sternotomy. All shunts were done with a Gore-Tex vascular graft (GTVG, W.L. Gore assoc. Inc, Elkton, MD). The most commonly used shunt size was 3.5mm (n=37), followed by 4mm (n=8), 3mm (n=1). Two out of three patients of the RV-PA conduit group were placed a 5mm graft, and the remaining patient a 7mm graft.

Group R, the early total correction group (n=15), underwent a VSD closure, and right ventricular outflow tract (RVOT) reconstruction, options of which included transannular patch (TAP, n=4), GTVG (n=5), bovine jugular vein graft (Contegra; Medtronic, Inc, Minneapolis, Minn, n=6), and all patients underwent routine resection of hypertrophied right ventricular (RV) muscle.

Statistical analysis

All data are presented as means or medians with standard deviations as well as frequencies accordingly. For continuous variables, we used the unpaired t test and the $\chi 2$ test for

categoric variables. Statistical significance was set at p value less than 0.05. The freedom from death or re-intervention including reoperations were analyzed using the Kaplan-Meier method and the Cox proportional Hazards Model. For the analysis of intergroup comparison of multiple events, the Prentice, Williams, and Peterson (PWP) model was used.

Results

Baseline Characteristics

Baseline characteristics of all patients are summarized in Table 1. The prevalence of situs inversus, left-sided aortic arch, juxtaductal stenosis (JDS) of the pulmonary arteries, and disconnected PA's did not differ between the two groups. Also, the type of VSD's (perimembranous, subarterial, muscular outlet, total conal defect) did not also differ significantly between the two groups. The pulmonary artery index, as analyzed by the Nakata method [2] showed a significant between the two groups; (Group R: 164.5±51.9 mm2/m2, Group P: 124.6±50.9 mm2/m2, p=0.010)

Table 1. Baseline Characteristics of the Two Groups

	Group P Mean (Range)	Group R Mean (Range)	*p*-value
Number of neonates under 90 days	49 (76.9%)	14 (23.1%)	
Age at first operation (days)	28.5 (9-77)	36.5 (6-90)	0.337
Birth Weight (kg. *)	2.64 (0.91-3.75)	2.85 (1.63-4.33)	0.272
Body Weight at first operation (kg.)	3.28 (2.5-4.5)	3.65 (2.4-5.8)	0.114

*kilograms

49 patients underwent palliative shunt (central, RMBT, LMBT, or RV-PA conduit), and 15 patients underwent initial total correction with various techniques (Rastelli-type repair with closure of the VSD and reconstruction of the RVOT with conduit or transannular patch).

Early Surgical Outcome & Intermediate Stage Mortality

Table 2. Early surgical outcomes

	Group P Mean (Range)	Group R Mean (Range)	*p*-value
Postoperative ventilator support (days)	3 (1-50)	6 (3-48)	0.095
Postoperative ICU* stay (days)	4 (1-50)	9 (3-48)	0.063
Postoperative hospital stay (days)	10 (1-50)	17 (4-48)	0.029
Operative Mortality (n, %)	4 (8%)	2 (13.3%)	

*Intensive Care Unit

Group R had a tendency towards longer postoperative ventilator support, and in the ICU (intensive care unit), and had a significant higher hospital stay than group R. In the R group,

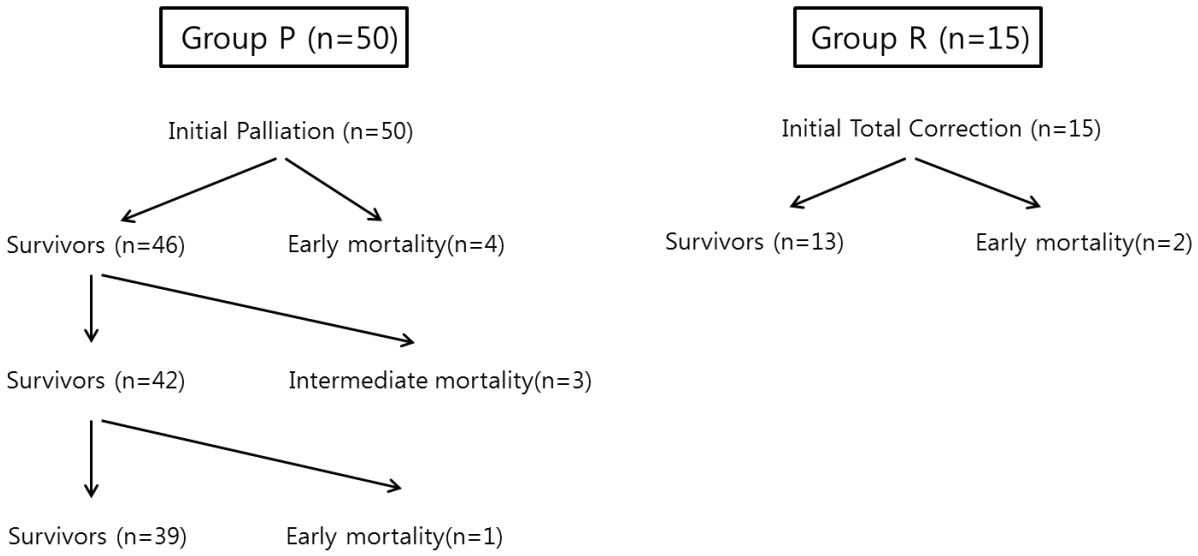
one patient showed low cardiac output following her initial total correction; hypoxic brain damage was complicated by hospital-acquired pneumonia which led to her death on postoperative day 97, and one other patient was discharged without any specific complications, but was lost in follow up due to death of unknown cause at home. In group P, two patients died due to pulmonary overflow that were not resuscitated, one due to a severe case of sepsis that did not respond to antibiotics therapy, and one other patient due to esophageal bleeding, onset by her previous surgery for congenital trachea-esophageal fistula.

There were also 3 cases of intermediate stage mortality; all three patients died at another hospital due to unknown cause, but perhaps due to shunt underflow caused by dehydration. A total of 46 patients in the P group underwent total correction later on, and 13 patients in group R were due to our follow-up after their total correction. There was one mortality in the P group in the total correction stage, an uneventful Rastelli-type repair was completed, however, the patient underwent cardiac arrest due to an unknown cause, and was put on extracorporeal membrane oxygenator (ECMO) for 45 days, but died due to multi-organ

failure. A flow chart of both groups is illustrated in Figure 1.

Fig 1. Postoperative course of both groups.

(Group P=Initial palliation Group, Group R=Initial total repair group)



Time-Related Survival after Birth

The 5-year survival rate for the entire cohort was 84.4%. As for group P, the 5-year survival rate was 86.7% for group R, and 83.6% for group P, which was statistically insignificant (p-value = 0.75). Most of the mortalities in the shunt group occurred in the palliation stage, including the immediate postoperative and intermediate stage.

Freedom from Death or Reinterventions

Reintervention was defined as surgical RV-PA conduit change, catheter interventions including ballooning, stenting, and outflow tract relief following their total correction. All instances of reinterventions were reviewed from medical records and categorized. A total 24 patients (36.9%) patients underwent at least one intervention, 10 patients (15.4%) underwent two, and 4 (6.2%) underwent three reinterventions. Of the total of 42 interventions, RV-PA conduit change was the most common with 19 cases (45.2%), followed by RVOT relief and PA ballooning/stenting at 10 cases each (23.8%), and 3 cases (7.1%) of surgical PA angioplasty.

The freedom from death or 1st reintervention at 5 years was 61% for group P, and 39% for group R with a significant inter-group difference (p=0.04) (Figure 2.). When analyzing the risk factors for reintervention using the cox proportional hazards model, Group (P or R) was the only significant risk factor (p =0.049). Group P showed a higher freedom from death or 2nd reoperations at 5 years than group R (Group P=77%, Group R=34%), with a significant inter-group difference (p=0.026). Also, Group was the only significant risk

factor for the 2nd reoperation as well (p=0.033). (Fig 2.) (Table 3.) All trends in occurrences of death or reinterventions were plotted chronologically for better visualization. (Fig 3.)

To further analyze the difference between the two groups in terms of death or any reintervention after birth, a Prentice, Williams, and Peterson (PWP) model of recurrent event model was applied. In both total time approach and gap time approach, Group P showed a significant advantage over Group R. (Table 4.)

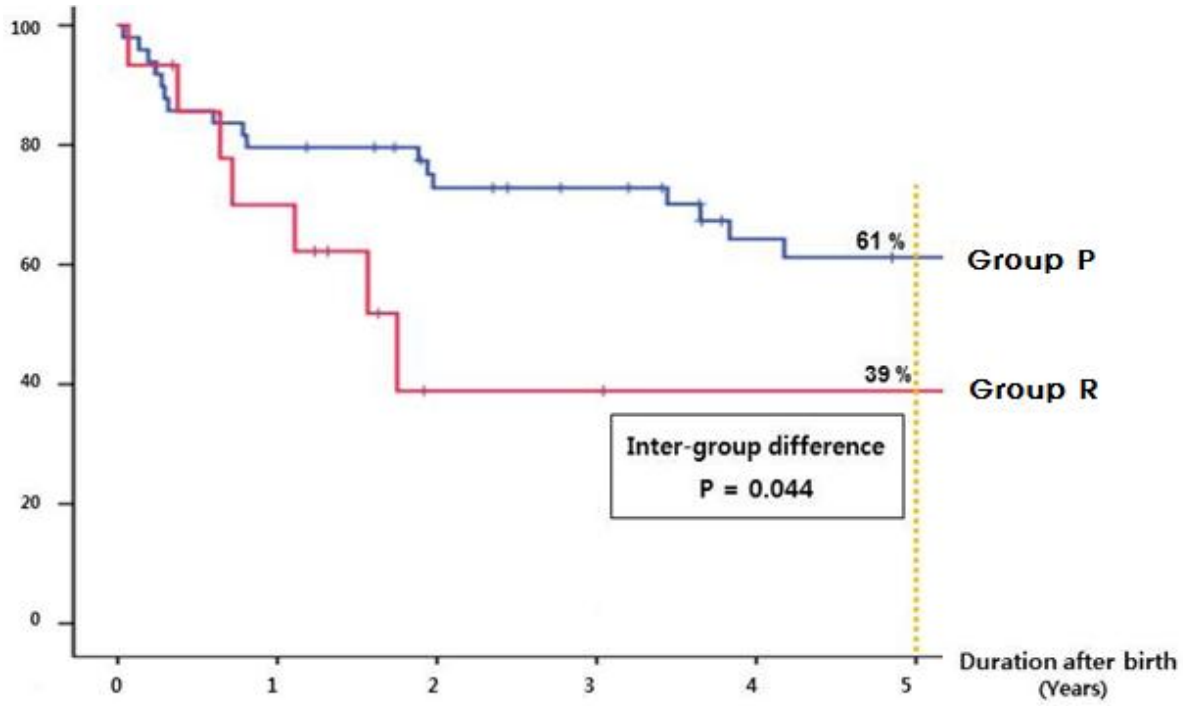
Table 3. Prentice, Williams, and Peterson (PWP) model of recurrent events.

	Hazard Ratio	95% CI	*P*-Value
Total Time Approach			
Group R	1		
Group P	0.2751	0.105-0.720	0.0086
Gap Time Approach			
Group R	1		
Group P	0.244	0.096-0.619	0.003

Fig 2. Time-dependent freedom from reoperation or death in the entire surviving cohort.

Freedom from death or 1st reintervention. (top)

Freedom from death or 2nd reintervention (bottom)



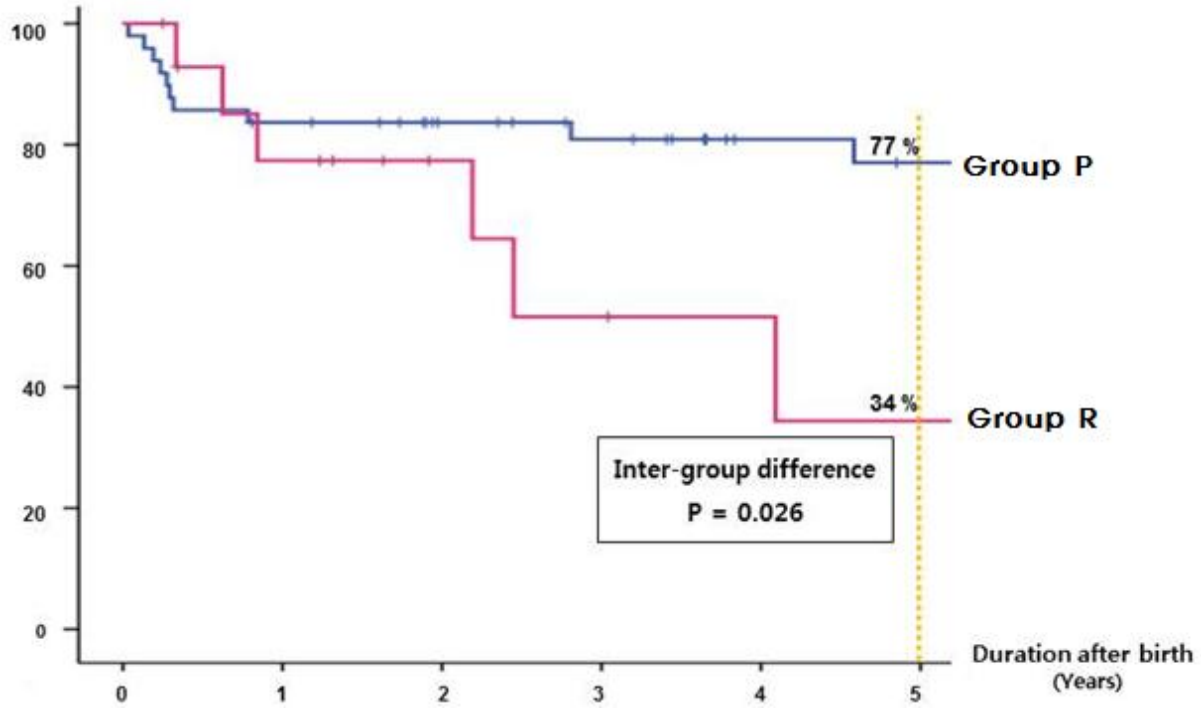


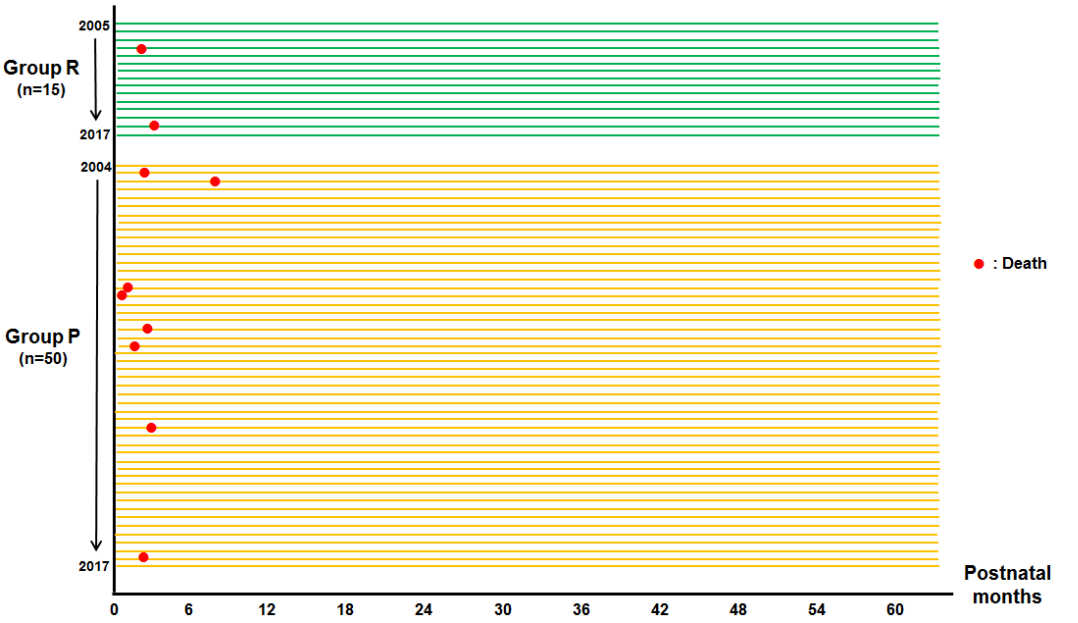
Table 4. Cox Proportional Hazards Model of 1st Reintervention or Death and 2nd Reintervention or Death

	Univariate Analysis	Multivariate Analysis		
	P-Value	Hazard Ratio	95% CI	*P*-Value
Group R against Group P (1st Reintervention or Death)	0.049	2.34	1.002-5.453	0.049
Group R against Group P (2nd Reintervention or Death)	0.033	2.91	1.090-7.752	0.033

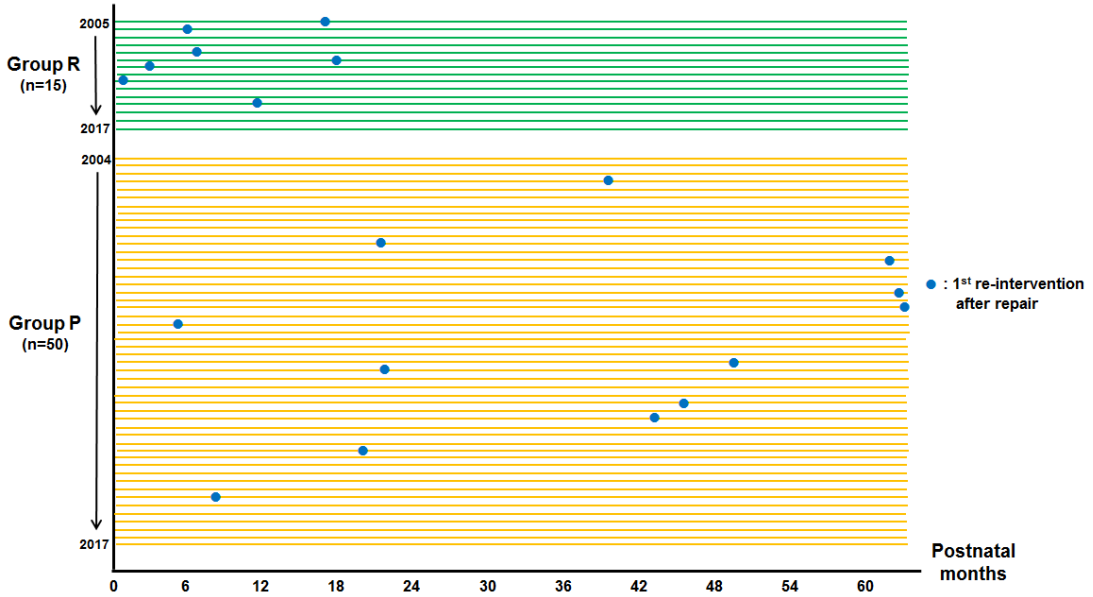
*CI-Confidence Interval

Fig 3. Chronological plotting of adverse events. (A) death (B) 1st reintervention (C) 2nd reintervention

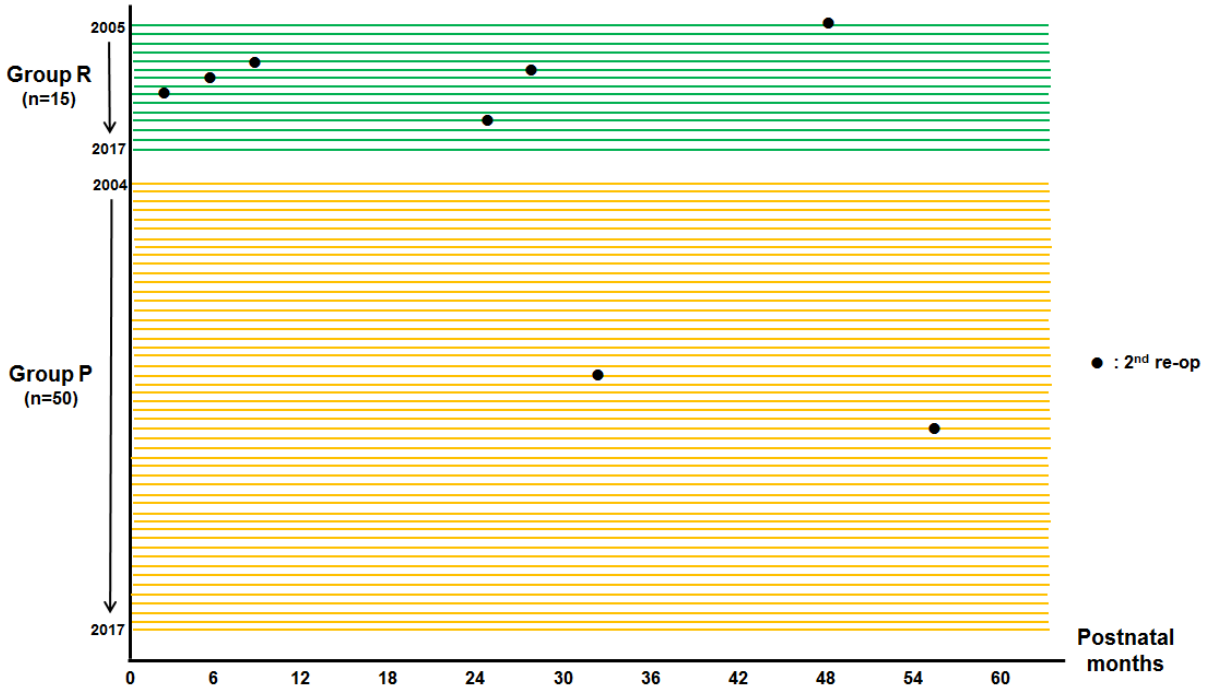
A)



B)



C)


2005
Group R
(n=15)
2017
2004
Group P
(n=50)
2017
● : 2nd re-op
0
6
12
18
24
30
36
42
48
54
60
Postnatal
months

Discussion

The initial choice of operation for complex congenital heart disease in the neonatal stage is still up to debate. In the spectrum of tetralogy of fallot (ToF) without pulmonary atresia, reported results of early repair have been excellent, providing evidence to the current consensus for elective repair after early infancy. [3] However, in symptomatic ToF patients, options of early repair versus palliative shunt followed by repair have been debated. [4] [5] [6] [7] In PAVSD patients, interstage mortalities, as well as operative mortalities following a palliative shunt have been mentioned as reasons why initial total correction may be a better option. [8][9] Furthermore, careful consideration is crucial for a shunt in ToF with pulmonary atresia because an occlusion or malfunction of any sort will eliminate any kind of forward pulmonary artery flow. To support this, a study by Alsoufi and colleagues in 2016 pointed out the anatomical setting of pulmonary atresia is a risk factor for mortality/morbidity in patients undergoing shunt palliation. [10] However, initial total correction entails the use of cardiopulmonary bypass in the neonatal stage, possible right ventricular dysfunction, and the inevitable placement of a relatively smaller RV-PA conduit.

[11] Although the possible detriments and possible advantages of each option have been argued, there have been limited studies comparing the two different surgical options, especially in terms of long-term rate of all sorts of reinterventions.

The outcomes of a palliative shunt have been of great interest for many years, with debatable outcomes. The Society of Thoracic Surgeons (STS) data published in 2017 of 9,172 infants showed in-hospital shunt failure in 674 (7.3%) patients. [12] Another retrospective study by Hobbes and colleagues in 2017 with 173 ToF / PAVSD with or without MAPCAs patients showed an in-hospital mortality of 5.2%, and inter-stage mortality of 3.6%. [13]

In our series, staged repair following a palliative shunt was associated with higher inter-stage mortality. However, as shown in figure 3, the occurrence of such events have decreased in recent years, which is attributed to advancements in ICU care, as well as a stringent home monitoring system at our institution, which without a doubt, has been established as a crucial factor. [14] [15] Parents of neonates discharged after a shunt operation are thoroughly educated, and are given a 24-hour hotline number for any signs of distress, which may

explain the decreasing occurrences of interstage mortalities in recent years. Also, we tend to minimize the interstage period by setting the timing of total correction within 6 months after birth, a point which we believe is appropriate in terms of minimizing interstage duration, as well as a long-lasting conduit placement. The study mentioned conducted by Hobbes and colleagues found that a high platelet count is associated with shunt failure, [11] highlighting the importance of appropriate anticoagulation needed to preserve shunt patency. After experiencing interstage mortalities due to shunt failure, we felt the need to increase the amount of acetylsalicylic acid, which was doubled to 10mg/kg, as compared to the previous 5mg/kg. It also must be noted that there has been a multicenter, multinational study to lower thrombotic risks in neonate and infants who underwent a palliative shunt, the CLARINET trial, which no benefit of additional clopidogrel. [16].

Another surgical issue that must be addressed is the presence of juxtaductal stenosis (JDS), which may be given careful thought in the management of PAVSD patients. [17] In specific, stenosis of the left pulmonary artery is a well-known, common cause of reoperation/reintervention in ToF patients. [18] A surgical protocol we apply at our

institution is the avoidance of direct manipulation of JDS, and we apply a BT shunt on the other side of the JDS, in order to allow and observe the gradual growth and native pulmonary artery tissue while ductal tissue will diminish. Then, insufficient size of the branch PA's will be surgically managed concomitantly at the time of total correction when ductal tissue is completely vanished. The reason behind our strategy is the inability to visually discern normal arterial tissue from ductal tissue in the presence of JDS.

We believe further advancements in ICU care, along with our current home monitoring program provided by experienced staff, the staged repair following a palliative shunt can gain a definite advantage over the initial total correction. In addition, although the shunt procedure is another surgery in itself, the longer patency and lower occurrences of reinterventions shown in our data can support such a statement.

We aimed to evaluate the outcomes of each surgical strategy in overall survival, freedom from reoperation, and pulmonary artery growth. We were not able to randomly assign neonates into two distinct operative strategies. However, they did not differ significantly in their baseline characteristics other than their PAIs, which was greater in the total correction

(group R) group, which somewhat decided their initial choice of operation.

Our study is limited in that this is a retrospective analysis, and presents a rather small number of patients undergoing initial total correction.

Conclusion

Staged repair strategy, as compared to initial total repair, was associated with higher inter-stage mortality with less frequent reinterventions after repair, which may be attributable to the use of larger conduits upon repair. Lowering the inter-stage mortality in the staged repair may allow for better surgical outcome in the future.

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국문요약

연구 배경: 신생아 및 영유아에서의 심실중격결손을 동반한 폐동맥폐쇄증에 대한 수술적인 치료는 단계적 교정술과 일차 완전 교정술이 될 수 있으나, 현재 어떤 방법이 단기적으로나 장기적으로 우수한지 정립되어 있지 않다. 이 것을 규명해보고자 연구를 시작하였다.

연구 방법: 2004 년부터 2017 년 까지 서울아산병원에서 심실중격결손을 동반한 폐동맥폐쇄증에 대하여 수술을 시행한 생후 90 일 미만의 아이 65 명을 의무기록으로 검색하여 분석하였다. 션트 수술 시행 이후 단계적 교정술을 시행한 아기 50 명, 그리고 일차 완전 교정술을 시행한 아기 15 명을 분석하였고, 생존율, 재수술/시술에 대한 빈도, 폐동맥의 성장 등을 분석하였다.

연구 결과: 전체 환자들의 중앙값 관찰 기간은 42.9 개월이였다. 두 그룹간의 수술 시 나이는 큰 차이가 없었다 (Group R: 36.5 ± 27.9 days, Group P: 28.4 ± 15.6 p=0.298), 체표면적 (Group R: 0.2 ± 0.0, Group P: 0.2±0.0, p=0.105), 하지만 폐동맥 지표 (나카타 인덱스) 에서는 유의한 차이가 있었다 (Group R: 164.5±51.9 mm2/m2, Group P: 124.6±50.9 mm2/m2, p=0.010). 전체 기간 중 9 례의 외과적인 사망이 발생 하였다

(Group P=7, Group R=2), 완전 교정술 이후 24 례의 첫 재수술/시술 (Group P=16, Group R=8), 그리고 11 례의 2 번째 재수술/시술 (Group P=3, Group R=8) 이 발생하였다. 5 년동안의 전체 생존율은 두 그룹 사이 큰 차이는 없었다. (group R=86.7%, group P=83.6%, p=0.754) (group R=86.7%, group P=83.6%, p=0.754), 하지만 단계적으로 완전 교정술을 받은 환아들은 재수술에 대한 위험이 낮았다 (HR 2.73, 95% CI:0.2751-0.7204, p=0.0086). 콕스 모델에서도 유일하게 의미 있게 나온 사망 및 첫 재수술/시술에 대한 위험 인자는 첫 수술의 형태였다 (HR 2.53, 95% CI:1.002-5.453; P=0.049), 사망 및 두 번째 재수술/시술에 대해서도 마찬가지 였다 (HR 2.90, 95% CI:1.09-7.75, p=0.026).

연구 결론: 단계적 완전 교정술은 일차적인 완전 교정술에 비하여 더 높은 중간 단계 사망률을 보이지만, 재수술/시술에 대한 위험도는 낮다. 이는 아마 완전 교정술을 할 때에 더 큰 도관이 들어가기 때문으로 생각되고, 미래에는 이런 중간 단계의 사망률을 낮추는 것이 중요할 것으로 사료된다.

중심 단어: 심실중격결손, 폐동맥폐쇄증