

# Surgical treatment of tetralogy of Fallot in symptomatic neonates and young infants



Bobae Jeon, MD, Dong-Hee Kim, MD, Bo Sang Kwon, MD, Eun Seok Choi, MD, Chun Soo Park, MD, PhD, and Tae-Jin Yun, MD, PhD

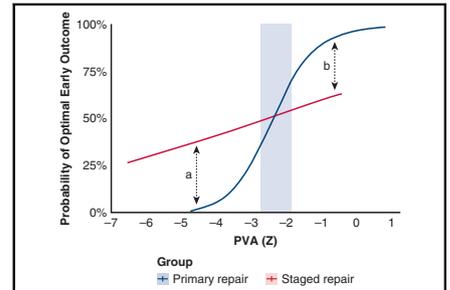
## ABSTRACT

**Objectives:** Optimal management of tetralogy of Fallot in symptomatic neonates and young infants remains controversial.

**Methods:** A retrospective review of 53 patients (26 male) with symptomatic tetralogy of Fallot who underwent primary repair (group 1, n = 22) or initial palliation (group 2, n = 31) within 2 months (60 days) after birth between 2005 and 2018 was performed. Subsequent repairs were performed in 29 patients at postpalliation 7.1 months in group 2 except for 2 interstage mortalities. Optimal early outcome was defined as no significant pulmonary stenosis or significant pulmonary regurgitation, and no reintervention within 12 months after repair.

**Results:** In group 2, median Z-score of the pulmonary valve annulus and McGoon ratio increased after palliation from -3.52 to -2.95 ( $P = .074$ ) and from 1.31 to 1.93 ( $P < .001$ ), respectively. Pulmonary annulus preservation at repair and optimal early outcome were achieved in 38 patients (17/22, 77%, group 1; 21/29, 72%, group 2) and 26 patients (12/22, 55%, group 1; 14/29, 48%, group 2), respectively. On logistic regression analysis, initial Z-score of the pulmonary valve annulus was the only predictor of annulus preservation at repair (odds ratio, 1.715,  $P = .0204$ ) and optimal early outcome (odds ratio, 1.583,  $P = .0259$ ). The annulus preservation probability curve according to the initial postnatal Z-score of the pulmonary valve annulus of all patients with repair (n = 51) showed an annulus preservation probability less than 70% in 3 patients (3/22) in group 1 and greater than 85% in 8 patients (8/29) in group 2, signifying that the alternative strategy might have been beneficial for each subset.

**Conclusions:** The surgical strategy for symptomatic tetralogy of Fallot should be individualized according to the initial size of the pulmonary valve annulus. (J Thorac Cardiovasc Surg 2020;159:1466-76)



Probability of OEO after repair in each group according to PVA (Z).

## CENTRAL MESSAGE

For symptomatic neonates and young infants with ToF, primary repair can be attempted in patients with a sizable PVA, but staged repair may be a better option in patients with a marginally small PVA.

## PERSPECTIVE

Optimal surgical strategy for symptomatic neonates and young infants with TOF is still under debate. Initial palliation with staged repair is associated with interstage mortality, whereas primary repair may lead to a higher incidence of TAP. Individualized surgical strategy for each patient based on PVA size may lead to optimal surgical outcomes.

See Commentaries on pages 1477 and 1478.

Optimal management of tetralogy of Fallot (TOF) in symptomatic neonates and young infants remains controversial. Primary repair with pulmonary annulus preservation (AP) may be an ideal option for patients with a sizable pulmonary

valve annulus (PVA), but patients with a marginally small PVA may benefit from initial palliation to allow for the growth of the PVA and increased probability of AP at later repair. Because a systemic-to-pulmonary shunt (SPS) for

From the Division of Pediatric Cardiac Surgery, Asan Medical Center, University of Ulsan College of Medicine, Seoul, Republic of Korea  
Institutional Review Board approval date: 2019.04.20. Institutional Review Board registration number: S2018-0707-0002

Read at the 99th Annual Meeting of The American Association for Thoracic Surgery, Toronto, Ontario, Canada, May 4-7, 2019.

Received for publication May 2, 2019; revisions received Oct 14, 2019; accepted for publication Oct 27, 2019; available ahead of print Jan 8, 2020.

Correspondence: Tae-Jin Yun, MD, PhD, Division of Pediatric Cardiac Surgery, Asan Medical Center, University of Ulsan College of Medicine, 88, Olympic-ro 43-gil, Songpa-gu, Seoul, Korea 05505 (E-mail: [tjyun@amc.seoul.kr](mailto:tjyun@amc.seoul.kr)).

0022-5223/\$36.00

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<https://doi.org/10.1016/j.jtcvs.2019.10.172>

**Abbreviations and Acronyms**

AP	= annulus preservation
CI	= confidence interval
CPB	= cardiopulmonary bypass
ICU	= intensive care unit
LPA	= left pulmonary artery
mBT	= modified Blalock–Taussig
OEO	= optimal early outcome
PS	= pulmonary stenosis
PR	= pulmonary regurgitation
P <sub>RV/LV</sub>	= ratio of the systolic pressure of the right ventricle to the left ventricle
PTFE	= polytetrafluoroethylene
PV	= pulmonary valve
PVA	= pulmonary valve annulus
PVA (Z)	= Z-score of the pulmonary valve annulus
RVOT	= right ventricular outflow tract
SPS	= systemic-to-pulmonary shunt
TAP	= transannular patch
TOF	= tetralogy of Fallot
VSD	= ventricular septal defect



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small babies (<2.5 kg) may lead to pulmonary overcirculation and hemodynamic instability, alternative palliative measures, or even primary repair, may be alternative options for this subset. However, multiorgan dysfunction, sepsis, genetic anomalies, severe prematurity, and other noncardiac conditions may preclude primary repair accompanied by cardiopulmonary bypass (CPB).<sup>1,2</sup> Therefore, the surgical strategy for symptomatic neonates and young infants should be delicately individualized, considering each patient's anatomic, demographic, and clinical characteristics. Given that the ultimate goal in ToF repair is leaving less pulmonary regurgitation (PR) and less pulmonary stenosis (PS) after repair,<sup>3</sup> the determination of optimal surgical strategy should center around the preservation of the structural integrity of the pulmonary valve (PV) (ie, AP) at repair.<sup>4</sup> In this study, we focused on ascertaining the optimal surgical strategy for each patient to achieve a higher incidence of AP at repair and satisfactory postrepair early outcomes.

**MATERIALS AND METHODS****Patients**

We performed a retrospective review of 53 symptomatic patients (26 male) with ToF who underwent surgical intervention within 2 months (60 days) after birth between 2005 and 2018. Patients with pulmonary atresia, ToF with absent PV syndrome, and ToF with atrioventricular septal defect were excluded from the study cohort. All patients showed persistently low (n = 36) or severely fluctuating (n = 17) oxygen saturation before the initial surgical intervention. The patients were divided into 2 groups according to their initial surgical strategy: primary repair (group 1, n = 22) and initial palliation (group 2, n = 31). Median age and body weight at initial surgical treatment, incidence of low birth weight and prematurity, and baseline oxygen saturation were comparable between the 2 groups, but initial postnatal PVA (Z) and McGoon ratio were significantly lower in group 2 (Table 1). Major associated anomalies were aortic valvular stenosis (n = 1),<sup>5</sup> infracardiac total anomalous pulmonary venous drainage (n = 1), aortopulmonary window (n = 1), vascular ring (n = 1) in group 1, and multiple rhabdomyoma (n = 1),<sup>6</sup> anomalous origin of the left pulmonary artery (LPA) from the ascending aorta (n = 1), LPA interruption (n = 1), and 2 major aortopulmonary collateral arteries with hypoplastic central pulmonary artery (n = 1) in group 2. Data collection, collation, and analysis were approved by the Institutional Review Board (No. S2018-0707-0002), and the need for informed consent was waived because of the retrospective nature of the study.

**Surgical Techniques**

Selection of palliative procedure in group 2 was based on the individual surgeon's preference, the patient's clinical conditions, and anatomic characteristics. Initial palliation comprised a modified Blalock–Taussig (mBT) shunt with (n = 14) or without (n = 13) CPB, right ventricular outflow tract (RVOT) widening (n = 2), hybrid RVOT stenting (n = 1), and placement of a right ventricle to pulmonary artery conduit (n = 1). For the mBT shunt, polytetrafluoroethylene (PTFE) vascular grafts (Gore-Tex expanded-PTFE Vascular Graft, WL Gore & Associates, Flagstaff, Ariz) of varying diameter (3 mm in 1 patient, 3.5 mm in 22 patients, 4 mm in 4 patients) were used. Sidedness of the mBT was central in 16 patients, right-sided in 9 patients, and left-sided in 2 patients. RVOT widening was performed for 2 small babies (body weight: 1 kg and 2.3 kg) who were deemed to have higher risks of pulmonary overcirculation and hemodynamic instability with an mBT shunt. A double-layered PTFE vascular patch (Gore Acuseal cardiovascular patch, WL Gore & Associates) was placed in the RVOT incision without pulmonary valvotomy to restrict the pulmonary blood flow at the valve level. Hybrid RVOT stenting was performed for a baby with a diminutive PVA that was deemed too small to be preserved later on at repair even if PVA outgrowth over the somatic growth could be achieved with the placement of an mBT shunt. Under CPB assistance, a Palmaz-Genesis stent (Cordis, Johnson and Johnson, Miami, Fla) was introduced through the main pulmonary arteriotomy, and the stent was balloon dilated to reach the final diameter and length of 5 mm and 15 mm, respectively. For right ventricle to pulmonary artery conduit implantation, a 4-mm expanded-PTFE vascular graft was placed under CPB assistance.

For the primary and staged repair of ToF, all patients underwent surgical correction with moderately hypothermic CPB and cardioplegic arrest, except for 4 patients with brief periods of deep hypothermic circulatory arrest. The ascending aorta was crossclamped and oblique right atriotomy was made to explore the ventricular septal defect (VSD) and RVOT through the tricuspid valve. Because surgical exposure of the VSD through the tricuspid valve was limited in neonates and young infants, the VSD was closed through the minimal right ventriculotomy (<10 mm) in most of the patients in group 1 (20/22, 91%). For the staged repair in group 2, the parietal extension of the infundibular septum was resected extensively through the tricuspid valve until the PV was clearly seen from the right

TABLE 1. Patient characteristics

Variable	Overall (n = 53)	Group 1 (n = 22)	Group 2 (n = 31)	P value
<b>Preoperative variables</b>				
Age, d	22 (5-59)	23 (6-58)	20 (5-59)	.398
Male sex	26/53 (49%)	11/22 (50%)	15/31 (48%)	.910
Birth weight, kg	3.04 (0.99-4.13)	3.05 (1.33-3.95)	3.01 (0.99-4.13)	.761
Low birth weight (<2.5 kg)	12/53 (23%)	4/22 (18%)	8/31 (26%)	.523
Gestational age, wk	38.6 (32.3-48.6)	38.7 (32.3-40.6)	38.6 (34.0-48.6)	.331
Prematurity (<37 wk)	9/53 (17%)	4/22 (18%)	5/31 (16%)	.848
Weight at initial surgery, kg	3.48 (1.0-4.43)	3.7 (2.50-4.43)	3.35 (1.0-4.4)	.510
Initial postnatal PVA (Z)	-2.70 (-6.56-0.7)	-2.25 (-4.71-0.7)	-3.71 (-6.56~-0.52)	.001
Initial postnatal McGoon ratio	1.36 (0.78-2.32)	1.41 (0.97-2.32)	1.31 (0.78-1.87)	.03
Initial baseline SpO <sub>2</sub> , %	60 (20-95)	65 (20-95)	50 (20-85)	.091
Age at repair, d	140 (6-834)	23 (6-58)	252 (75-834)	<.001
Weight at repair, kg	6.8 (2.5-11.0)	3.7 (2.50-4.43)	7.7 (5.4-11.0)	<.001
Pre-repair PVA (Z)	-2.40 (-9.21-1.27)	-2.25 (-4.71-0.7)	-3.01 (-9.21-1.27)	.001
Pre-repair McGoon ratio	1.64 (0.97-2.86)	1.41 (0.97-2.32)	1.92 (1.1-2.86)	.02
<b>Operative variables</b>				
Emergency or urgent operation	45/53 (85%)	19/22 (86%)	26 (26/31, 84%)	.702
PVA preservation at repair	38/51 (75%)	17/22 (77%)	21/29 (72%)	.458
Type of valve-sparing repair				
Isolated infundibular patch	5/38 (13%)	4/17 (24%)	1/21 (5%)	
Infundibular + supra-valvar patch	31/38 (82%)	12/17 (70%)	19/21 (90%)	
Isolated supra-valvar patch	2/38 (5%)	1/17 (6%)	1/21 (5%)	
Transatrial VSD closure	11/51 (22%)	2/22 (9%)	9/29 (31%)	.05
CPB time (min)	137 (74-231)	106.5 (74-231)	143 (104-212)	.075
Aortic crossclamp time, min	65 (36-143)	64 (44-143)	66 (36-125)	.128
<b>Postoperative variables</b>				
Mechanical ventilation, d	2 (0.3-80)	6 (0.6-80)	0.75 (0.3-8)	.011
Postoperative ICU, d	5 (1-92)	12 (1-92)	2 (1-14)	.002
Hospital length of stay, d	10 (5-117)	19.5 (5-117)	8 (6-30)	.006
Mortality	2/53 (4%)	0/22 (0%)	2/31 (6%)	.233

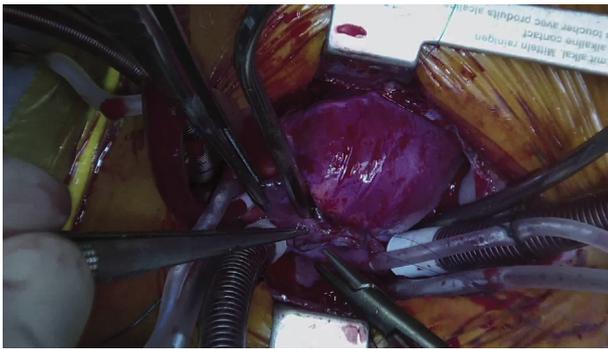
PVA (Z), Z-score of pulmonary valve annulus; SpO<sub>2</sub>, saturation of percutaneous oxygen; VSD, ventricular septal defect; CPB, cardiopulmonary bypass; ICU, intensive care unit.

ventricular inlet. A Hegar dilator was introduced through the tricuspid valve to measure the PV orifice diameter. If the orifice was larger than a normal PVA minus 5 mm, we elected not to explore the PV through the pulmonary arteriotomy. We referred to echocardiography-based nomograms to ascertain the normal PVA diameter of each patient.<sup>7</sup> If the PV orifice was smaller than the normal PVA minus 5 mm,<sup>8</sup> the main pulmonary artery was incised longitudinally for the careful assessment of PV morphology. If the valve leaflets were thin and functional, the PV was left intact without any intervention for AP. Otherwise, various surgical techniques, including commissurotomy, commissural mobilization by excising the web-like structure around the commissures, and shaving of the lumpy valve leaflets, were used to increase the effective PV orifice area. PVA size was then measured again with Hegar dilators. If it was still smaller than normal PVA dimension minus 5, the pulmonary arteriotomy was extended down to the RVOT, crossing the annulus (<10 mm) for limited transannular patch (TAP) placement. After the patient was weaned from CPB, the ratio of right ventricular pressure to left ventricular pressure (P<sub>RV/LV</sub>) and the pressure gradient between the right ventricle and the pulmonary artery were directly measured. If right ventriculotomy had not been performed and the P<sub>RV/LV</sub> was greater than 0.8 with a significant gradient between the right ventricle and the main pulmonary artery, the patient was put back on CPB and a mini-infundibular incision (10 mm) was made for RVOT muscle resection and infundibular patch placement using an Acuseal patch. If the P<sub>RV/LV</sub> was still higher than 0.8 after

infundibular patch placement, the patient went back on CPB for TAP. Various peripheral pulmonary angioplasty techniques (eg, patch angioplasty, carinoplasty in the bifurcation of the main pulmonary artery, and left pulmonary artery wedge resection and repair to correct the acute angulation and stenosis) were used if indicated (Video 1).

**Surgical Outcomes**

The outcomes of interest were all-cause death (identified by medical records), PVAP at repair, major complications within 30 days after surgery, and development of significant PS (RVOT flow velocity ≥ 3.5 m/s) and PR (moderate or more). Major complications were defined as unplanned reoperation, arrhythmia requiring permanent pacemaker implantation, cardiac arrest, circulatory instability requiring mechanical support (extracorporeal membrane oxygenation or ventricular assist device), acute renal failure requiring hemodialysis or hemofiltration, neurologic deficit persisting at discharge, phrenic nerve palsy with or without diaphragmatic plication, and deep wound infection or mediastinitis. To ascertain the individualized surgical approach based on preoperative anatomical and demographic characteristics, patients with optimal early outcome (OEO) were identified. OEO was defined as the absence of any of the following events during the first year after repair: death (all-cause), RVOT obstruction (peak velocity ≥ 3.5 m/s), PR (grade moderate or more), and reintervention (catheter-based or surgical) after repair.



**VIDEO 1.** Emergency surgical intervention for ToF was performed for a 23-day-old girl with repeated episodes of cyanotic spell. Her body weight and PVA (Z) at operation were 4.1 kg and  $-2.0$ , respectively. For better surgical exposure of VSD and complete relief of infundibular stenosis, a small right ventriculotomy (10 mm in length) was made. After the parietal band was extensively excised, the large perimembranous VSD was closed through the right ventriculotomy. A longitudinal incision was made on the main pulmonary artery to explore the PV morphology, which showed severe commissural fusion of the right commissure. PV orifice diameter was initially measured as 3 mm by Hegar dilator insertion through the right ventriculotomy before PV intervention and was enlarged to 5 mm after extensive commissurotomy, which was 4.1 mm smaller than normal (9.1 mm). Both pulmonary arteriotomy and right ventriculotomy were closed with round Acuseal patches. CPB time and aortic crossclamping time were 88 minutes and 57 minutes, respectively. Postrepair right ventricle to left ventricle pressure ratio was 0.65 (43 mm Hg/66 mm Hg). On intraoperative echocardiography, there was trivial PR and no VSD leakage, and RVOT flow velocity was 3.0 m/sec. He was discharged without any complication on postoperative day 9. Follow-up echocardiography at postoperative 8 months showed RVOT flow velocity of 2.8 m/sec with mild PR. Video available at: [https://www.jtcvs.org/article/S0022-5223\(19\)33088-0/fulltext](https://www.jtcvs.org/article/S0022-5223(19)33088-0/fulltext).

### Statistical Analysis

Categorical variables were presented as frequencies and percentages, and continuous variables were presented as mean with standard deviation or median with interquartile range according to the distribution of the data. Distributional normality was tested using the Kolmogorov–Smirnov method. Kaplan–Meier survival estimation was used for the analysis of time-related adverse events, and differences between the subgroups were tested using the log-rank test. For the matched comparison of the outcomes between the 2 groups, patients in each group were matched by age at repair, birth weight, and PVA (Z). To identify the risk factors for the decreased time to composite events of significant PS (ie, RVOT flow velocity  $\geq 3.5$  m/s) or PR (moderate or more) after repair, Cox proportional hazards model was fitted. Predictors of AP at repair and OEO were identified using logistic regression analysis, and probability curves for AP and OEO according to the variables of interest were plotted to identify subsets in each group who would have benefited from alternative surgical strategies.

Statistical analysis was conducted with SPSS Statistics version 22 (IBM, Armonk, NY), R software version 3.4.4 ([www.r-project.org](http://www.r-project.org)), and GraphPad statistical software package version 5 (GraphPad, San Diego, Calif).

### RESULTS

There was 1 early death and 1 late death, both in group 2. A full-term female baby with a birth weight of 3.3 kg who underwent emergency central shunt procedure under CPB assist with a 3.5-mm PTFE vascular graft at postnatal day

5 developed necrotizing enterocolitis at 5.5 months postoperatively and died of sepsis. Another premature baby (34<sup>+</sup> gestational age) weighing 990 g who underwent RVOT patch widening under CPB assist at postnatal day 10 could not come off CPB and died of low cardiac output on postoperative day 1. All 8 patients with major associated anomalies (ie, intracardiac total anomalous pulmonary venous drainage, aortopulmonary window) survived. The only major complication in group 1 was an incident of cardiac arrest on the day of operation, and the patient recovered without significant sequelae. Open sternum with delayed sternal closure was performed in 13 patients in group 1 (13/22, 59.1%). Follow-up was complete in all patients, and the median follow-up duration was 62.3 months (interquartile range, 16.5–104.4 months). The 5-year survivals of group 1 and group 2 were 100% and 93.5%  $\pm$  4.4%, respectively (Figure E1). In group 2, the median PVA (Z) and the McGoon ratio increased after palliation from  $-3.52$  to  $-2.95$  ( $P = .074$ ) and from 1.31 to 1.93 ( $P < .001$ ), respectively. Except for the 2 interstage mortalities, subsequent repairs were performed in the remaining 29 patients at 7.1 months after palliation in group 2. There was no perioperative death in group 2 after repair, and major complications included reoperation for VSD leakage in 1 patient, delayed sternal closure in 3 patients, and diaphragmatic plication for phrenic nerve palsy in 2 patients. Operative and postoperative characteristics are summarized in Table 1.

### Total Length of Stay and Medical Expenditure

Postrepair mechanical ventilation time was significantly longer in group 1 than in group 2 (Table 1), and the former was even longer than the summation of postpalliation and postrepair mechanical ventilation time in group 2 (group 1: median 6 days, 0.6~80 days; group 2: median 4 days, 1.5~21.4 days,  $P = .043$ ). Although postrepair intensive care unit (ICU) stay and hospital stay in group 1 were significantly longer than those in group 2 (Table 1), they were comparable to the summations of postpalliation and postrepair ICU stay and hospital stay in group 2: ICU stay (group 1: median 12 days, 1~92 days; group 2: 10 days, 4~28 days,  $P = .148$ ), hospital stay (group 1: median 20 days, 5~117 days; group 2: 22 days, 13~64 days,  $P = .301$ ). Total medical expenditure for primary repair in group 1 was comparable to the summation of the medical expenditure for palliation and repair in group 2 (group 1: median: 22,964 USD, range: 8727–63,385 USD, group 2: median 23,388 USD, range: 9813–59,560 USD,  $P = .516$ ).

### Annulus Preservation Probability

AP was achieved at repair in 38 patients (17/22, 77%, in group 1, 21/29, 72%, in group 2,  $P = .458$ ). On logistic regression analysis, initial postnatal PVA (Z) was identified as the only predictor of AP at repair (odds ratio, 1.715 per 1 increase of PVA (Z), 95% CI, 1.087–2.705,  $P = .0204$ ,

**TABLE 2. Logistic regression analysis for the probability of annulus preservation**

Variable	Univariate <i>P</i> value	Multivariate <i>P</i> value	OR	Lower 95% CI	Upper 95% CI
Sex	.1994				
Age at initial operation	.5355				
Birth weight	.3732				
Gestational age	.1926				
Initial postnatal PVA (Z)* (per 1 increase)	.0083	.0204	1.715	1.087	2.705
Initial McGoon ratio†	.1496				
Body weight at initial operation	.2749				
Surgical strategy	.6933				
Type of palliation	1.0000				
Body weight at repair	.6748				

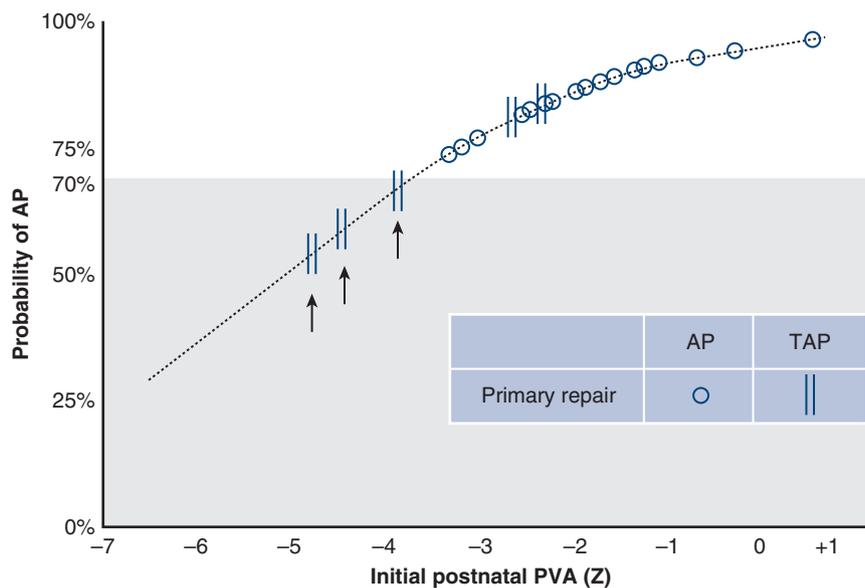
OR, Odds ratio; CI, confidence interval; PVA (Z), Z-score of pulmonary valve annulus. \*Preoperative PVA (Z) was measured by echocardiography and calculated with an echocardiographic nomogram. †Preoperative McGoon ratio was measured by echocardiography and computed tomography. Variables with a univariate *P* value below .2 were used for multivariate analysis.

**Table 2).** Based on the AP probability curve by the initial PVA (Z) of the patients who underwent repair (n = 51), AP probability was lower than 70% in 3 patients (3/22) in group 1 (Figure 1) and higher than 85% in 8 patients (8/29) in group 2 (Figure 2), which signified that the alternative strategy for each subset might have been beneficial in terms of AP probability. The median AP probability of a subset with initial postnatal PVA (Z) between -4.5 and -2.5 in group 2 increased from 68% (range, 59%-82%) to 81% (range, 42%-92%) (*P* = .3). Freedom from significant postrepair PS (55.7% ± 12.2% in group 1 and 71.2% ± 8.6% in group 2, *P* = .39, Figure E2), significant postrepair PR (42.8% ± 10.6% in

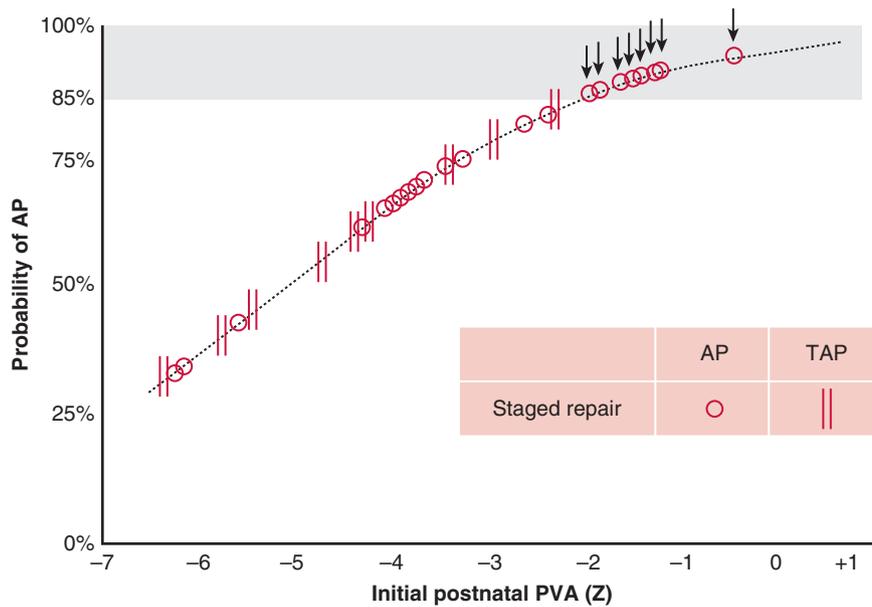
group 1 and 50.6% ± 13.9% in group 2, *P* = .86, Figure E3), and postrepair reintervention for PS or PR (57.1% ± 12.9% in group 1 and 69.9% ± 9.0% in group 2, *P* = .21, Figure E4) at 5 years were comparable between the 2 groups.

**Matched Comparison of the Outcomes Between the 2 Groups**

Patients in each group were matched by age at repair, birth weight, and PVA (Z), and 21 matched pairs (total 42 patients) were identified. Overall survival (100% in group 1 and 95.2% ± 4.7% in group 2, *P* = .33, Figure E5), freedom from significant postrepair PS (58.3% ± 12.5%



**FIGURE 1.** Depiction of AP probability according to the initial postnatal PVA (Z) of each patient with primary repair shows that the alternative surgical strategy (ie, staged repair) might have been better in terms of increasing the probability of AP for 3 patients whose AP probability based on initial PVA (Z) was lower than 70% (arrows). AP, Annulus preservation; PVA (Z), Z-score of the pulmonary valve annulus; TAP, transannular patch.



**FIGURE 2.** Depiction of AP probability according to the initial postnatal PVA (Z) in each patient with initial palliation shows that the alternative surgical strategy (ie, primary repair) might have been better in terms of avoiding unnecessary palliation for 8 patients whose AP probability based on initial PVA (Z) was higher than 85% (arrows). AP, Annulus preservation; PVA (Z), Z-score of the pulmonary valve annulus; TAP, transannular patch.

in group 1 and 75.0% ± 9.7% in group 2,  $P = .49$ , [Figure E6](#)), freedom from significant postrepair PR (50.6% ± 13.9% in group 1 and 34.0% ± 11.5% in group 2,  $P = .63$ , [Figure E7](#)), and freedom from postrepair reinterventions for PS or PR (52.1% ± 12.8% in group 1 and 53.2% ± 11.5% in group 2,  $P = .85$ , [Figure E8](#)) at 5 years were comparable between the 2 groups.

**Cox Regression for Decreased Time to Pulmonary Stenosis or Regurgitation**

Initial PVA (Z) was identified as the only risk factor for decreased time to composite outcomes of significant PS or PR (hazard ratio, 0.75; 95% confidence interval [CI],

0.563-1.000;  $P = .05$ ), and surgical strategy was not a significant risk factor for decreased time to the development of the composite events of significant PS or PR ( $P = .74$ ).

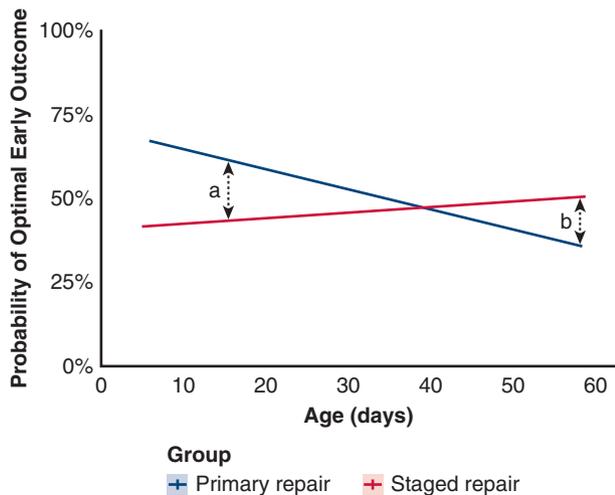
**Optimal Early Outcome**

OEO was achieved in 26 patients (group 1 = 12, group 2 = 14). On logistic regression analysis, initial postnatal PVA (Z) was identified as the only predictor of OEO (odds ratio, 1.583 per 1 increase of PVA (Z), 95% CI, 1.057-2.371,  $P = .0259$ , [Table 3](#)). The OEO probability curve of the patients who completed repair (n = 51) by the age ([Figure 3](#)) at initial operation, body weight

**TABLE 3. Logistic regression analysis for the predictors of optimal early outcome**

Variable	Univariate P value	Multivariate P value	OR	Lower 95% CI	Upper 95% CI
Sex	.0745				
Age at initial operation	.7314				
Birth weight	.1363				
Gestational age	.6725				
Initial postnatal PVA (Z)* (per 1 increase)	.0124	.0259	1.583	1.057	2.371
Preoperative McGoon†	.7115				
Body weight at initial operation	.0889				
Surgical strategy	.5007				
Type of palliation	.7054				
Body weight at repair	.1843				

OR, Odds ratio; CI, confidence interval; PVA (Z), Z-score of pulmonary valve annulus. \*Preoperative PVA (Z) was measured by echocardiography and calculated with an echocardiographic nomogram. †Preoperative McGoon ratio was measured by echocardiography and computed tomography. Variables with a univariate P value below .2 were used for multivariate analysis.

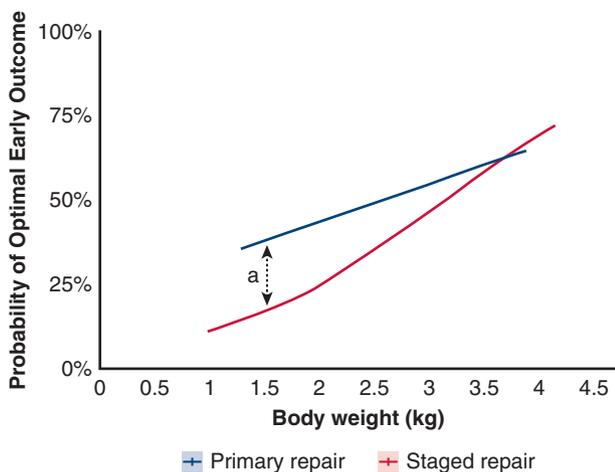


**FIGURE 3.** Depiction of the probability of OEO according to the age at initial surgical intervention in the 2 groups shows that the primary repair strategy may be beneficial for younger patients (gap a) and the staged repair strategy may be better for older patients (gap b) in terms of achieving OEOs.

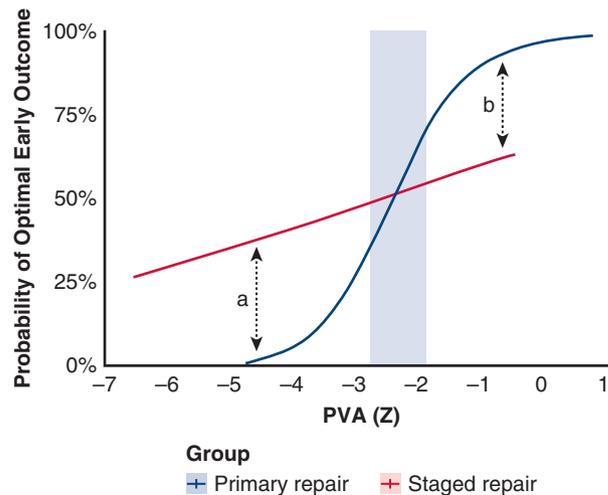
(Figure 4) at initial operation, and initial postnatal PVA (Z) (Figure 5) showed that the estimated probability of achieving OEO was higher with the primary repair strategy in younger and smaller patients with larger PAV (Z), whereas the probability was higher with the staged repair strategy in older and larger patients with smaller PAV (Z).

**DISCUSSION**

Although early outcomes after the elective repair of ToF are excellent in the contemporary series, the optimal surgical management for symptomatic neonates and young



**FIGURE 4.** Depiction of the probability of OEO according to the body weight at initial surgical intervention in the 2 groups shows that the primary repair strategy may be beneficial for smaller patients (gap a) in terms of achieving OEOs.



**FIGURE 5.** Depiction of the probability of OEO according to the initial PVA (Z) in the 2 groups shows that the staged repair strategy may be beneficial for patients with smaller PVA (Z) (gap a) and the primary repair strategy may be better for patients with larger PVA (Z) (gap b) in terms of achieving OEOs. PVA (Z), Z-score of the pulmonary valve annulus.

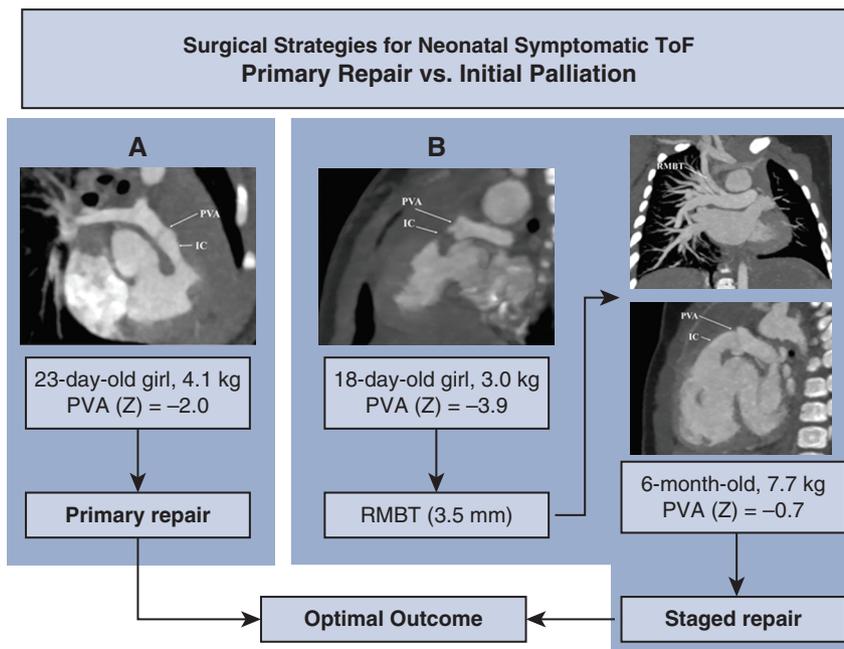
infants remains to be defined. Primary repair appears to be preferred in this setting,<sup>1,9</sup> but placement of SPS is still the procedure of choice in many programs.<sup>4,10-12</sup> In a multicenter analysis enrolling 342 patients with ToF who were registered in the Society of Thoracic Surgeons database after undergoing various neonatal surgical interventions, initial palliative procedures were more frequently performed than repair (ie, palliation in 178 patients and primary repair in 152 patients).<sup>10</sup> The ratio of palliation over repair may be even higher if the number of elective repair procedures for acyanotic and asymptomatic neonates based on an institutional bias toward routine early primary repair<sup>13-15</sup> is excluded.

Current indications for an SPS in patients with ToF are (1) severely hypoplastic pulmonary arteries; (2) extracardiac conditions precluding primary repair, such as sepsis, viral respiratory infection, intracranial hemorrhage, and other organ dysfunction<sup>1</sup>; and (3) a marginally small PVA.<sup>4,16</sup> SPS may facilitate the growth of the PVA, and SPS-induced outgrowth of the PVA over somatic growth may lead to a higher probability of PVA preservation upon staged repair in patients with a marginally small PVA who, with the primary repair strategy, would undergo a placement of a TAP.<sup>4,11,12,16,17</sup> In a study including 216 infants with TOF, of whom 29 infants initially underwent SPS with a subsequent repair and 187 infants received a primary repair, the PVA (Z) increased significantly after the placement of an SPS ( $P = .001$ ), whereas the prerepair changes in the PVA (Z) in the primary repair group were not statistically significant ( $P = .7$ ), with a significant intergroup difference ( $P < .001$ ).<sup>16</sup> Because of the collinearity of the RVOT dimensions,<sup>17,18</sup> neonates and

young infants with profound hypoxemia attributable to severe infundibular stenosis tend to have a marginally small PVA, necessitating a TAP. This could well be the reason why previous reports pertaining to the outcomes after primary repair for symptomatic neonates with ToF have shown a consistently high incidence of TAP, ranging from 66% to 100%.<sup>9,11,15,19-22</sup> However, patients with a sizable PVA whose hypoxemia is due only to isolated infundibular stenosis do not necessarily need a TAP. Primary repair with PVA preservation and extensive infundibular widening may be the better surgical option for these patients (Figure 6).

With respect to the comparison of the outcomes after initial palliation and primary repair, a number of comparative studies have been conducted in the context of early and late mortality,<sup>23,24</sup> RVOT management at repair,<sup>21</sup> ICU stay and hospital stay,<sup>2,23,24</sup> and medical expenses and reintervention rate.<sup>25,26</sup> In this study, the frequencies of PVA preservation at repair in groups 1 and 2 were comparable (77% in group 1 vs 72% in group 2), even if initial postnatal PVA (Z) was significantly lower in group 2. Although there are several reports asserting that long-term adverse effects of isolated right ventricular incision with regard to reintervention or mortality are comparable to those of TAP,<sup>27,28</sup> it is generally accepted that preservation of the PVA per se, even at the expense

of infundibular incision<sup>29</sup> and leaving certain degree of RVOT gradient,<sup>30,31</sup> may be beneficial for the prevention of right ventricular dilatation necessitating PV implantation. If the placement of an SPS promotes the outgrowth of the PVA over the somatic growth, patients with a marginally small PVA could benefit from initial palliation in terms of increasing the probability of PVA preservation.<sup>4,12,16,17</sup> However, unadjusted comparison between the staged repair group and primary repair group in terms of the incidence of PVA preservation is usually inappropriate because the 2 groups are different in terms of RVOT dimensions. Furthermore, PVA preservation may not necessarily connote the best surgical outcome.<sup>27</sup> Because the ultimate goal in the treatment of ToF is leaving less PS and less PR in the long run,<sup>3</sup> we coined the acronym OEO, which was defined as “no mortality, no significant PS, no significant PR, and no reintervention dedicated to the RVOT obstruction at 1-year follow-up after the repair.” As expected, PVA size was identified as the only determinant predicting optimal surgical outcome. When the probability curves for OEO in relation to PVA (Z) were compared between the 2 groups, staged repair strategy was found to be superior to primary repair strategy in achieving better OEO in patients with smaller PVA, or vice versa, which signified that if the PVA size is marginally small, surgical outcome would be better with a staged repair strategy in that the probability



**FIGURE 6.** A, A 23-day-old girl with ToF (PVA-Z = -2.0) developed severe cyanosis and underwent primary repair with PVA preservation. Follow-up echocardiography at postoperative 8 months showed RVOT flow velocity of 2.8 m/s with mild PR. B, An 18-day-old girl with ToF (PVA-Z = -3.9) developed repeated episodes of hypoxic spell and underwent RMBT using a 3.5-mm PTFE vascular graft. Follow-up cardiac CT scan at postnatal 6 months showed significant increases in the size of PVA (PVA-Z = -0.7) and branch pulmonary artery, and staged repair with PVA preservation was performed. Follow-up echocardiography at postoperative 1 year showed RVOT flow velocity of 2.9 m/s with mild PR. *ToF*, Tetralogy of Fallot; *PVA*, pulmonary valve annulus; *PVA-Z*, Z-score of pulmonary valve annulus; *RMBT*, right modified Blalock-Taussig shunt.

of AP may increase after palliation, and if the PVA size is large enough, optimal surgical outcome is achievable without superfluous palliative measures. Therefore, the surgical strategy for symptomatic neonates and young infants should be individualized in terms of maximizing the probability of PVA preservation based on the anatomic disposition of each patient.

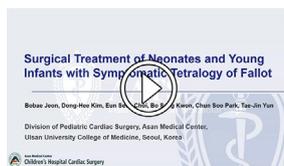
Various palliative procedures other than SPS have been adopted for symptomatic neonates and young infants with ToF, including infundibular patching or palliative TAP, right ventricle to pulmonary artery conduit implantation,<sup>32</sup> PV ballooning, and RVOT stenting.<sup>33</sup> Infundibular patching in patients with a sizable PVA or overly large TAP may lead to excessive pulmonary flow and hemodynamic instability. Right ventricle to pulmonary artery conduit placement requires right ventricular incision, which could be avoided with staged repair after the placement of an SPS. Because PV ballooning or RVOT stenting leads to the partial or complete destruction of the structural integrity of the PV, these procedures should be adopted with caution.

## CONCLUSIONS

A staged repair strategy is more frequently used than primary repair for symptomatic neonates and young infants with ToF. The surgical strategy for symptomatic ToF should be individualized according to the initial size of the PVA. Primary repair should be attempted for patients with a sizable PVA (Z), but staged repair may be a better option for patients with a marginally small PVA to increase the probability of AP. The estimated probability of achieving OEO was higher with the primary repair strategy in younger and smaller patients with larger PVA (Z), whereas the probability was higher with the staged repair strategy in older and larger patients with smaller PVA (Z).

## Webcast

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## Conflict of Interest Statement

Authors have nothing to disclose with regard to commercial support.

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**Key Words:** neonate, pulmonary valve annulus preservation, tetralogy of Fallot, transannular patch

## Discussion



**Dr Robert Jaquiss (Dallas, Tex).** This was a group of consecutive small infants, and I think we need to talk about the subpopulation. When we are talking about repair in everybody and preserving the valve in everybody, we need to know what age we are working with. This was the toughest, youngest, smallest age. These were profoundly cyanotic infants, and they were consecutive. The average oxygen saturations was approximately 55%, and yet in this tough group you were able to achieve annulus-sparing repair in more than 70%. This is a remarkable achievement.

With this approach, more than 50% of infants had an OEO using a stringent definition of optimal: no significant RVOT obstruction, no pulmonary insufficiency, and no reintervention in the first year. That's a terrific result. By 5 years with longer follow-up, the proportion of both cohorts was significant, residual RVOT obstruction was stable, but an increasing proportion had developed some

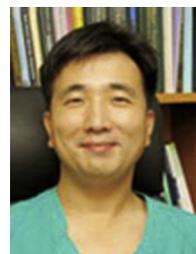
degree of significant pulmonary insufficiency. By 5 years, a little less than half in each group required reintervention, although some of those were clearly in the pulmonary artery and not in the outflow tract. The only variable that was predictive of a less than optimal outcome was the initial PV Z score.

You suggested in your presentation toward the end that a few patients in each group were perhaps inappropriately assigned; some people were palliated who didn't need it, some people who underwent early repair perhaps would have better been treated by initial palliation. Because this was a retrospective study, has your center now adopted a formal prospective treatment protocol for assignment of patients to appropriate treatment based on Z score, which looks to be approximately -2 to -3? Have you adopted a protocol to manage these patients or is it still determined on a case-by-case basis?

(No response)

**Dr Jaquiss.** Well, let's say yes and move on to the next question.

The proportion of patients with an early residual RVOT gradient of at least 50 mm Hg is somewhat high in your cohort, and I noticed from your description of your intraoperative approach that you used an right ventricle/left ventricle pressure ratio of 0.8. For anything higher than that, you went back, but that is rather higher than some people would say is ideal for revision threshold. Do you believe that your policy of tolerating a right ventricle/left ventricle pressure ratio of 0.8 is too permissive or do you believe that your high rate of AP justifies such an approach? Are you still doing the same thing?



**Dr Tae-Jin Yun (Seoul, Republic of Korea).** Regarding the first question, our institutional strategy is to repair ToF at approximately 3 or 4 months of age. We don't do neonatal elective repair. For the patients with a relatively sizable PVA, we would try to perform a repair with preserving the annulus at the neonatal period. If the PVA is marginally small, we would like to take advantage of staged repair strategy recently.

The second question is about the right ventricle/left ventricle pressure ratio.  $P_{RV/LV}$  of 0.8 might look a little bit higher than the criteria by others, but 0.8 is still the most frequently used cut-off value.  $P_{RV/LV}$  of patients in this study was usually approximately 0.5, which translates to right ventricle systolic pressure of 30 to 40 mm Hg. With respect to the late development of RVOT obstruction, it seems like that the patients who show high  $P_{RV/LV}$  intraoperatively do not necessarily develop RVOT obstruction.

**Dr Jaquiss.** My last question is one of timing just as a practical matter. How long should we wait after we have

done an initial shunt? Do you schedule elective repair in a palliated patient at 6 months or 5 months or 7 months or are you watching the PVA Z score? How do you know when to go back and do a complete repair?

**Dr Yun.** Well, it depends, because in some patients the outgrowth of the PVA is slow and very fast in others. So it depends on the growth of the PVA. The average age at repair is approximately 6 months in this study, including the staged repair group. If we look at the repair age of the primary repair group, it is only 3 or 4 months of age. Thus, repair age in the staged repair group was between 6 and 12 months.



**Dr James A. Quintessenza** (*Lexington, Ky*). I have another question about the right ventricle/left ventricle ratio of 0.8. Do you have a sense of postoperative interventional like balloon therapy, how often that is performed, and has that been effective in maintaining those valves?

**Dr Yun.** If we count all the reoperations, the freedom from reoperation is low in this study. However, when we actually look at the patients with reoperation dedicated to a small PVA, there were only 5 patients who had AP at repair and were converted to TAP, and there were another 5 patients who developed RVOT obstruction and were treated with postoperative ballooning of the PV, which turned out to be effective without causing significant PR. So it may be an elegant strategy to preserve the annulus as aggressively as possible and then do some catheter intervention later on, if necessary.

**Dr Quintessenza.** I think the 73% valve preservation with postoperative ballooning may be a reasonable strategy for this group.



**Dr Shunji Sano** (*Okayama, Japan*). I have a brief comment. From my experience of 25 years in Okayama, we have done more than 400 cases and no deaths, 1 late death, and then 60% of the patients had valve preservation and 20% had a mini-TAP. So the 20% is the more than

5 mm Hg to the right ventricle, and I use a TAP. The operation in 25 years is 13 patients. And 18% of the tetralogy have a shunt, and the timing of the total repair is usually 6 to 12 months after the initial palliation according to the valve size. The other indication was only an institutional problem. I have a maximum 200 waiting list, so I

couldn't do the operation in a timing, because I have only 6 ICU in the 400 cases. But the result is not much difference. We try aggressively to preserve the valve, and we use like a (inaudible) technique for more than 20 years.

**Unidentified Speaker.** In the 0.8 ratio that you accept, does it make a difference whether the majority of the pressure gradient is at the annular level or if there is a mix of infundibular residual dynamic narrowing and annular obstruction.

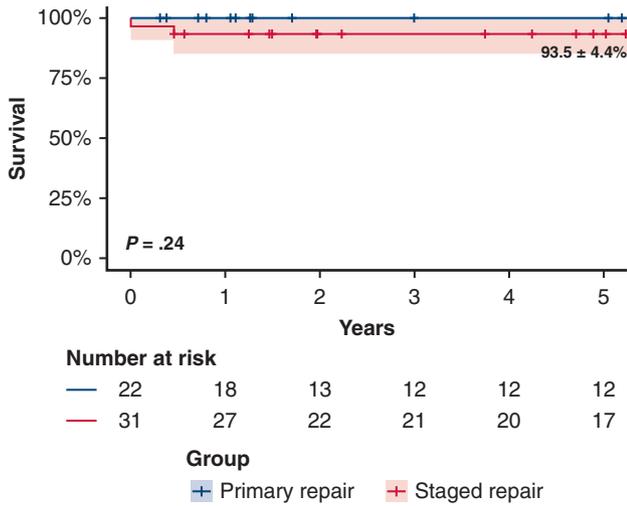
**Dr Yun.** To preserve the annulus in this challenging subset of patients, I think it is important to eliminate all the gradient below and above the valve. Because we tried to get rid of all the pressure gradient below the valve, RVOT obstruction was mainly due to the stenosis at the valve level.

**Unidentified Speaker.** There is a lot of dynamic obstruction in the RVOT immediately in the postoperative transesophageal echocardiography. Is this 0.8 also at the discharge echocardiogram, and what is the progression of it in the first midterm follow-up by the echo? What are you comfortable with in terms of the right ventricle/left ventricle ratio in the first 6 months?

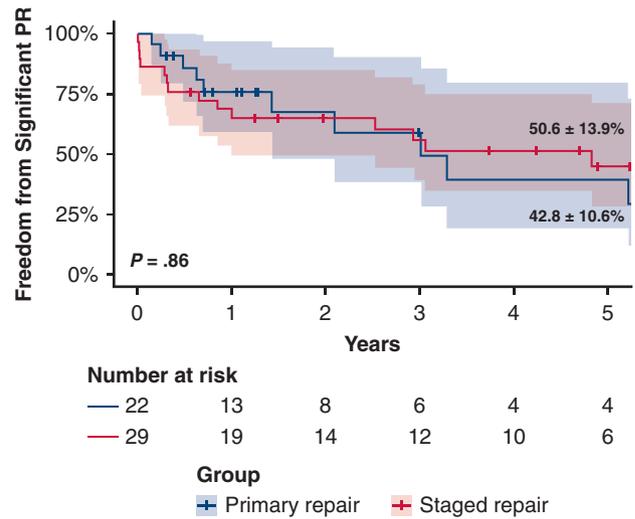
**Dr Yun.** Indication of the catheterization for the patients with significant RVOT gradient was RVOT flow velocity greater than 3.5 m/s on echocardiography, but we found that half of the patients did not have high right ventricle pressure in the catheterization laboratory even though the RVOT gradient by echocardiography was deemed significant. RVOT pressure gradients assessment by echocardiography is sometimes overestimating. Because we eliminate all the gradient below and above the PV, it's not dynamic obstruction. Some patients may develop valvular stenosis afterwards because of the inadequate catch-up growth of the PV, and postoperative balloon dilatation of the PV may be a viable option for those patients.

**Unidentified Speaker.** So as long as you achieve a good surgical result intraoperatively, your postoperative transesophageal echocardiography is not really a decision maker in terms of leaving the operating room or discharging the patient from the hospital?

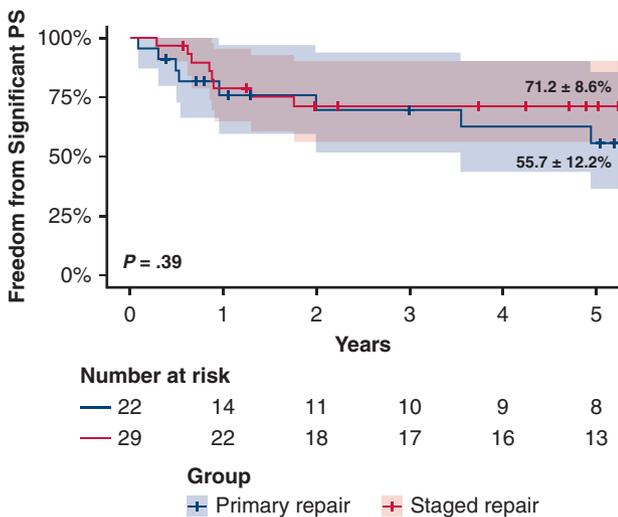
**Dr Yun.** We measure the right ventricle pressure directly, and we see the intraoperative transesophageal echocardiography. If  $P_{RV/LV}$  is higher than 0.8 by direct measurement or if the intraoperative transesophageal echocardiography shows that the flow velocity is more than 3.5 m/s, we would be worried about the residual RVOT obstruction, and we may put the patient back on bypass to place a TAP.



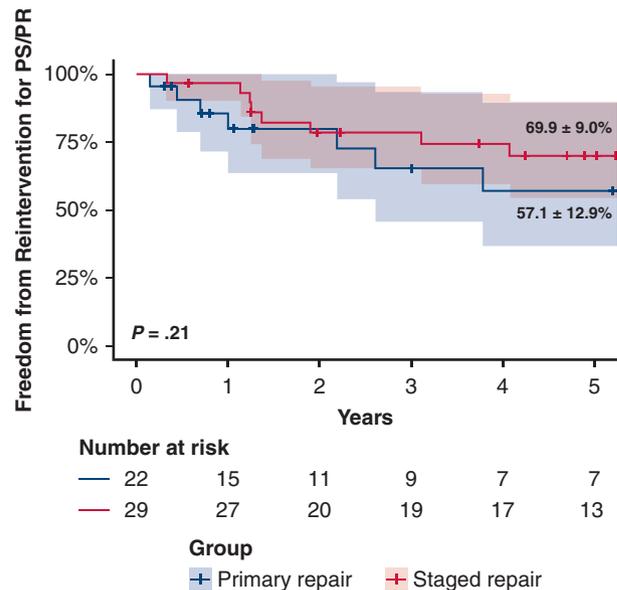
**FIGURE E1.** Kaplan-Meier survival estimate of the 2 groups shows no significant difference in overall survival ( $P = .24$ ).



**FIGURE E3.** Depiction of the freedom from significant PR (moderate or more) shows no significant difference between the 2 groups ( $P = .86$ ). PR, Pulmonary regurgitation.

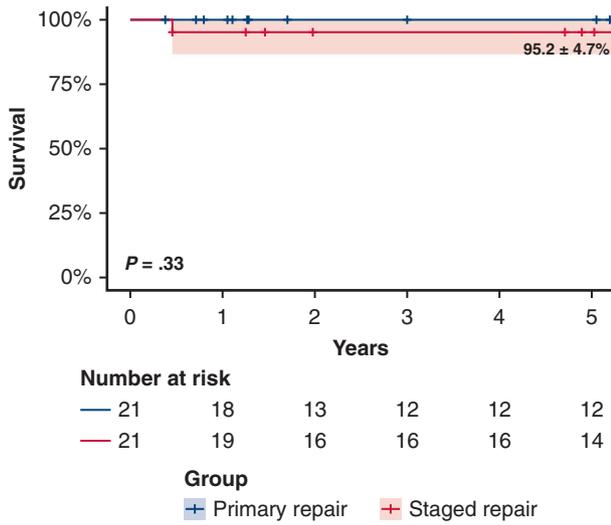


**FIGURE E2.** Depiction of the freedom from significant PS (RVOT flow velocity  $\geq 3.5$  m/s) shows no significant difference between the 2 groups ( $P = .39$ ). PS, Pulmonary stenosis.

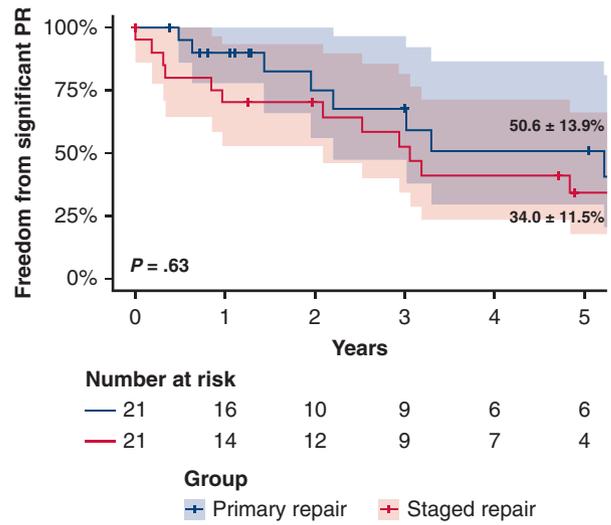


**FIGURE E4.** Depiction of the freedom from any surgical or catheter reintervention for PS or PR shows no significant difference between the 2 groups ( $P = .56$ ). PS, Pulmonary stenosis; PR, pulmonary regurgitation.

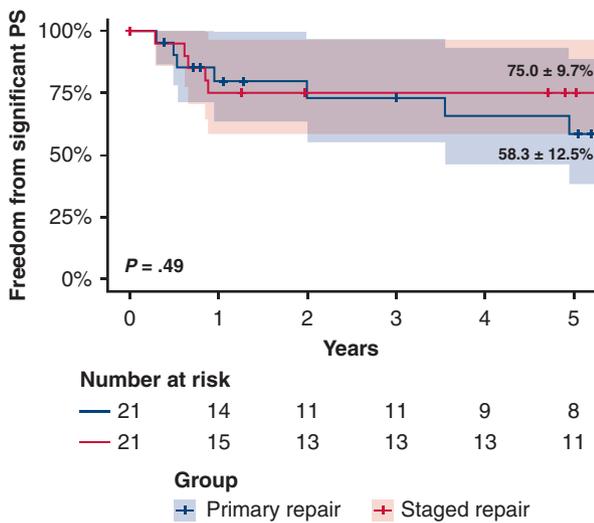
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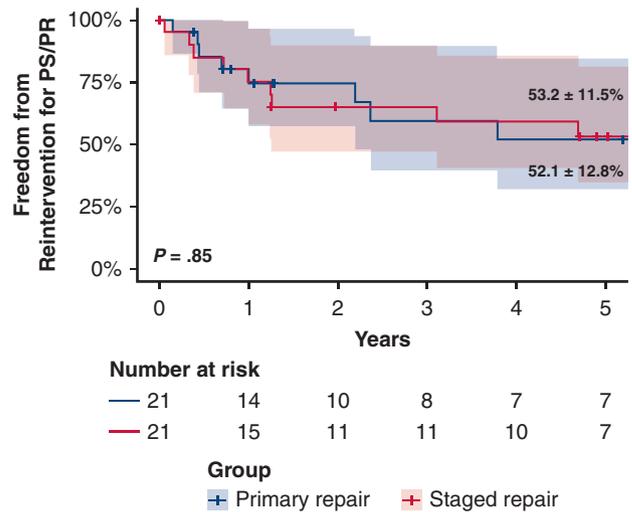
**FIGURE E5.** Kaplan–Meier survival estimate of the 21 matched pairs from each group shows no significant intergroup difference in overall survival ( $P = .33$ ).



**FIGURE E7.** Depiction of the freedom from significant PR (moderate or more) in the 21 matched pairs from each group shows no significant difference between the 2 groups ( $P = .63$ ). PR, Pulmonary regurgitation.



**FIGURE E6.** Depiction of the freedom from significant PS (RVOT flow velocity  $\geq 3.5$  m/s) in the 21 matched pairs from each group shows no significant difference between the 2 groups ( $P = .49$ ). PS, Pulmonary stenosis.



**FIGURE E8.** Depiction of the freedom from any surgical or catheter reintervention for PS or PR in the 21 matched pairs from each group shows no significant difference between the 2 groups ( $P = .85$ ). PS, Pulmonary stenosis; PR, pulmonary regurgitation.