JCS Journal of Chest Surgery
https://doi.org/10.5090/jcs.22.106
pISSN: 2765-1606 eISSN: 2765-1614
J Chest Surg. 2023;56(2):75-86

Clinical Research

Outcomes of Surgical Repair for Truncus Arteriosus: A 30-Year Single-Center Experience

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ARTICLE INFO
Received September 27, 2022
Revised November 14, 2022
Accepted November 29, 2022

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[†]This article was presented at the 54th annual meeting of The Korean Society for Thoracic and Cardiovascular Surgery in 2022.

See Commentary page 87.

Background: We investigated the long-term outcomes of truncus arteriosus repair at a single institution with a 30-year study period.
Methods: Patients who underwent repair of truncus arteriosus between 1993 and 2022 were reviewed retrospectively. Factors associated with early mortality, overall attrition, and reintervention were identified using appropriate statistical methods.
Results: In total, 42 patients were enrolled in this study. The median age and weight at repair were 26 days and 3.5 kg, respectively. Thirty patients (71.4%) underwent 1-stage repair. There were 8 early deaths (19%). In the univariable analysis, undergoing surgery before 2011 was associated with early mortality (p=0.031). The overall survival rate at 10 years was 73.8%. In the multivariable analysis, significant truncal valve (TrV) dysfunction (p=0.010), longer cardiopulmonary bypass time (p=0.018), and the earlier era of surgery (p=0.004) were identified as risk factors for overall mortality. During follow-up, 47 reinterventions were required in 27 patients (64.3%). The freedom from all-cause reintervention rate at 10 years was 23.6%. In the multivariable analysis, associated arch obstruction (p<0.001) and significant TrV dysfunction (p=0.011) were identified as risk factors for all-cause reintervention. Arch obstruction (p=0.027) and a number of TrV cusps other than 3 (p=0.014) were identified as risk factors for right ventricle to pulmonary artery (RV-PA) reintervention, and significant TrV dysfunction was identified as a risk factor for TrV reintervention (p=0.002).
Conclusion: Despite recent improvements in survival outcomes after repair of truncus arteriosus, RV-PA or TrV reinterventions were required in a significant number of patients during follow-up.

Keywords: Truncus arteriosus, Risk factors, Reintervention, Long-term outcome

Introduction

Truncus arteriosus (TA) is a rare congenital heart defect, the prevalence of which ranges from 60 to 107 per million live births [1,2]. In TA, a single truncal artery arises from the heart through a single truncal valve (TrV), where both aorta and pulmonary arteries originate. Other types of cardiovascular anomalies, including ventricular septal defect, TrV abnormalities, arch obstruction, and coronary artery anomalies, are frequently associated with TA [3,4]. At most centers, TA is repaired within the first few weeks of life [4-7], unless pulmonary obstructive disease, which is always life-threatening, could develop [4]. Although a staged approach has been adopted to avoid significant morbidity and mortality following neonatal surgery [8], early 1-stage repair has been a standard approach for this anomaly. With progress in surgical techniques and perioperative management, outcomes have been gradually improved [5,8,9]. While various factors associated with mortality and reinterventions have been identified in previous studies, mortality, morbidity, and reinterventions after repair are still not uncommon [5-7,9-12].

Therefore, the objective of this study was to investigate the perioperative and long-term outcomes after the repair of TA and to identify factors associated with the outcomes.

https://doi.org/10.5090/jcs.22.106

Methods

Patient selection and data collection

This study was approved by the Institutional Review Board of Asan Medical Center (IRB approval no., 2022-1105) and the requirement for informed consent was waived because of the retrospective nature of this study. All patients who underwent complete repair of TA between 1993 and 2022 were enrolled in this study. Baseline characteristics, operative details, perioperative outcomes, mortality, reinterventions, and follow-up data were collected by reviewing medical records and telephone contact.

Definitions

TA was categorized using the classification system developed by Collett and Edwards [13]. Type 4, with the lung supplied by collaterals, was not considered to belong to TA and excluded.

Significant TrV dysfunction included either significant TrV regurgitation (greater than mild degree) or significant

Table 1. Baseline characteristics of the patients

Characteristic	Value
Age at operation (day)	26 (9–256)
Neonate at operation	24 (57.1)
Prematurity	3 (7.1)
Weight at operation (kg)	3.50 (3.2–5.5)
Low weight at operation (<3.0 kg)	9 (20.9)
Sex	
Male	26 (61.9)
Female	16 (38.1)
Truncus arteriosus type	
I	27 (64.3)
II	11 (26.2)
III	4 (9.5)
No. of TrV cusps	
2	7 (16.7)
3	24 (57.1)
4	11 (26.2)
TrV regurgitation	
None	10 (23.8)
Mild	25 (59.5)
Moderate	5 (11.9)
Severe	2 (4.8)
TrV stenosis	
None	19 (44.2)
Mild	12 (27.9)
Moderate	9 (20.9)
Severe	2 (4.7)
Associated cardiac anomalies	
Ventricular septal defect	42 (100.0)
Atrial septal defect	31 (73.8)
Patent ductus arteriosus	7 (16.7)
Arch obstruction	6 (14.3)
Interrupted aortic arch	5 (11.9)
Coarctation of the aorta	1 (2.4)
Right aortic arch	4 (9.5)
Partial anomalous pulmonary venous return	3 (7.1)
Coronary anomaly (single coronary artery)	1 (2.4)
Chromosomal or noncardiac anomalies	10 (19.0)
DiGeorge syndrome/22q11.2 microdeletion	6 (14.3)
2p22.2 duplication	1 (2.4)
VATER syndrome	1 (2.4)
Others	2 (4.8)

Values are presented as median (interquartile range) or number (%).
TrV, truncal valve.

Table 2. Operative details

Variable	Value
Surgical strategy	
One-stage	30 (71.4)
Staged	12 (28.6)
Type of palliative surgery	
Truncal separation	6 (14.3)
Pulmonary artery banding	4 (9.5)
Right modified Blalock-Taussig shunt	1 (2.4)
Right ventricle to pulmonary artery connection	
Conduit	37 (88.1)
PTFE valved	11 (26.2)
Bovine jugular vein valved (Contegra)	10 (23.8)
Homograft	8 (28.6)
PTFE valveless	6 (14.3)
Others	3 (7.2)
Direct connection (réparation à l'étage ventriculaire)	5 (11.9)
Conduit diameter (mm)	12 (10–14)
Concomitant truncal valve surgery	7 (16.7)
Repair	5 (11.9)
Replacement	2 (4.8)
Concomitant cardiac surgery	
ASD closure	21 (50.0)
Arch repair	6 (14.3)
Patent ductus arteriosus ligation/division	2 (4.8)
Mitral valve repair	2 (4.8)
Tricuspid annuloplasty	1 (2.4)
ASD creation	1 (2.4)
Partial anomalous pulmonary venous return repair	1 (2.4)
Cardiopulmonary bypass time (min)	186 (139–236)
Aortic cross-clamp time (min)	84 (60–108)
Era	
2011 or later	22 (52.4)
Before 2011	20 (47.6)

Values are presented as number (%) or median (interquartile range).
PTFE, polytetrafluoroethylene; ASD, atrial septal defect.

TrV stenosis (greater than mild degree) assessed with transthoracic echocardiography. Significant TrV regurgitation was defined as a vena contracta width >3 mm and aortic diastolic flow reversal, and significant TrV stenosis was defined as aortic jet velocity >3 m/sec and a mean gradient >20 mm Hg, based on the recommendations for echocardiographic assessment proposed by the American Society of Echocardiography [14,15].

Early mortality was defined as death occurring within 30 days after the operation or before hospital discharge. Reintervention was defined as any procedure performed in the catheter lab or operating theater late after repair of TA. The study period was arbitrarily divided into 2 eras (earlier era, from 1993 to 2010; later era, from 2011 to 2022) to evenly distribute the number of patients; 20 patients were included in the earlier era and 22 patients in the later era.

The outcomes of interest were death or transplantation, all-cause reintervention, reintervention for connection between the right ventricle and pulmonary artery (RV-PA), and TrV reintervention.

Surgical strategy

One-stage repair has been our preferred repair strategy throughout the study period. However, in patients with medical conditions not suitable for the use of cardiopulmonary bypass, pulmonary artery banding (or bilateral pulmonary artery banding) was performed as an initial surgical intervention. When the branch pulmonary arteries were considered too small for complete repair, establishing forward flow from the right ventricle following truncal separation without repair of the ventricular septal defect could be considered to enhance branch pulmonary arterial growth for future complete repair. The decision to perform concomitant TrV repair was made based on the severity of dysfunction, reparability, and the individual surgeon's discretion.

Table 3. Factors associated with early mortality (n=8)

Variable	Odds ratio (95% confidence interval)	p-value
Sex		
Male	Reference	
Female	0.48 (0.08–2.71)	0.403
Prematurity	11.00 (0.86–141.33)	0.066
Age at operation (day)	1.00 (1.00–1.00)	0.480
Low weight at operation (<3.0 kg)	2.80 (0.52–15.04)	0.230
Truncus arteriosus type		
I	Reference	.
II	0.98 (0.16–6.00)	0.981
III	1.47 (0.12–17.21)	0.761
No. of TrV cusps		
3	Reference	
2 or 4	2.69 (0.55–13.20)	0.222
Significant TrV dysfunction[a)]	2.40 (0.50–11.54)	0.274
Arch obstruction	2.50 (0.37–16.89)	0.347
Chromosomal or noncardiac anomaly	1.08 (0.18–6.46)	0.930
Surgical strategy		
One-stage	Reference	
Staged	0.30 (0.33–2.73)	0.285
Type of right ventricle to pulmonary artery connection		
Conduit	Reference	
Réparation à l'étage ventriculaire	3.44 (0.47–25.23)	0.223
Concomitant TrV surgery	0.67 (0.07–6.47)	0.727
Cardiopulmonary bypass time	1.01 (1.00–1.02)	0.103
Aortic cross-clamp time	1.01 (0.99–1.03)	0.438
Era		
2011 or later	Reference	
Before 2011	11.31 (1.24–102.72)	0.031

TrV, truncal valve.
[a)]Significant TrV dysfunction was defined as truncal stenosis or regurgitation greater than mild.

https://doi.org/10.5090/jcs.22.106

Surgical technique

After a midline sternal incision was made, cardiopulmonary bypass was established with aorto-bicaval cannulation. The truncal artery was cross-clamped, and the truncal artery was opened and transected to evaluate the pulmonary arteries and coronary arteries. The truncal artery was carefully separated, and the TrV was addressed if needed. The truncal artery was repaired directly or using a patch. A longitudinal incision was made on the RV below the truncal root. The ventricular septal defect was closed with a patch through the RV incision. Finally, the RV-PA connection was accomplished, and the atrial septal defect could be completely closed or left open depending on RV function and pulmonary valve competence. The sternum could be left open if needed (n=17, 40.5%).

Statistical analysis

Categorical variables are described as frequencies and percentages, and continuous variables are described as the mean with standard deviation or the median with interquartile range according to the distribution of the data. The distributional normality of the data was tested using the Kolmogorov-Smirnov method. Differences between the groups were evaluated using the t-test or Wilcoxon signed rank test for continuous variables and the chi-square test or Fisher exact test for categorical variables, as appropriate. The analysis of overall survival or freedom from reintervention was conducted using the Kaplan-Meier method, and intergroup equality of survival curves was assessed using the log-rank test. Logistic regression analysis was conducted to determine the factors associated with early mortality after TA repair. A Cox proportional-hazards regression model was fit to identify independent risk factors associated with time-dependent events, including deaths or transplants, all-cause reinterventions, RV-PA reinterventions, and TrV reinterventions. A p-value of <0.05 was considered to indicate statistical significance. All analyses were performed using IBM SPSS statistical software ver. 26.0 (IBM Corp., Armonk, NY, USA).

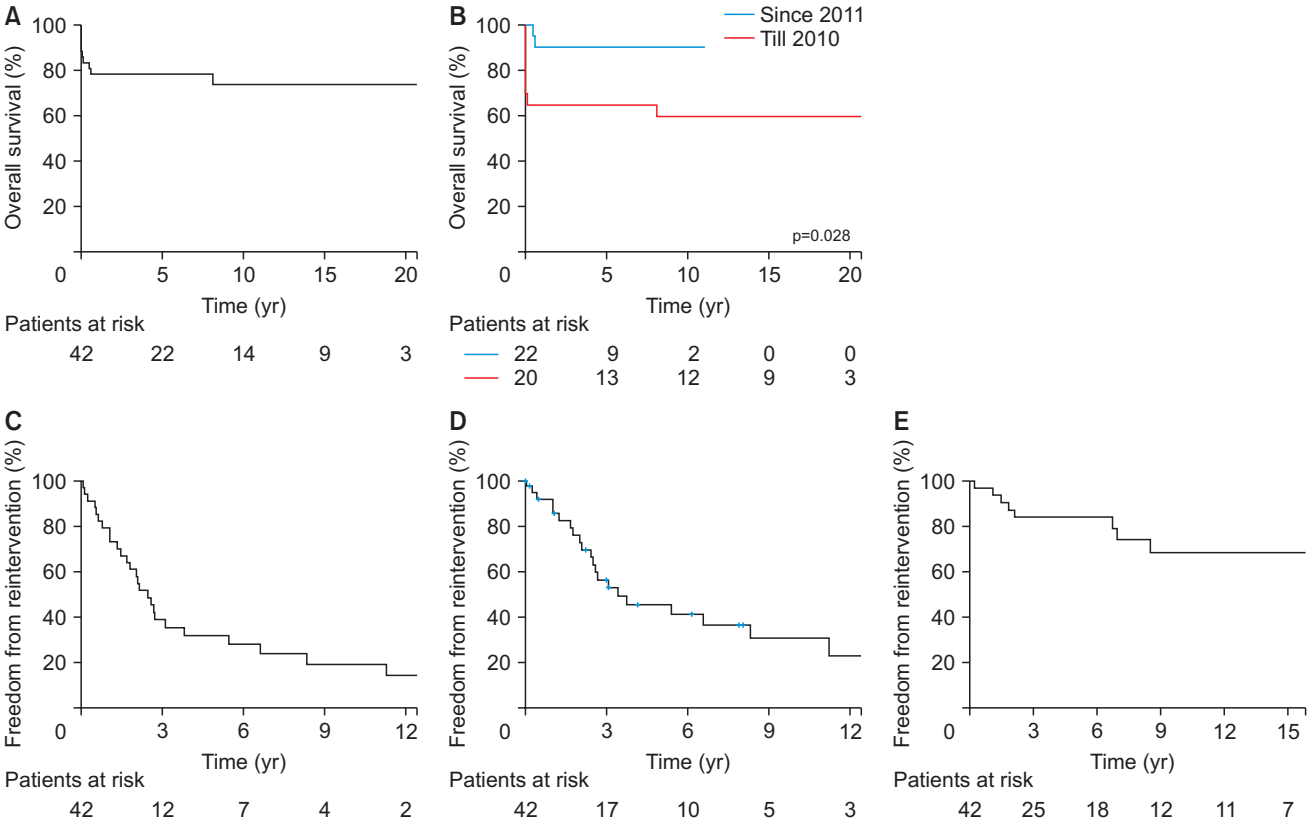

Fig. 1. Kaplan-Meier curves for survival and reintervention. (A) Overall survival. (B) Overall survival according to the era of surgery. (C) Freedom from all-cause reintervention. (D) Freedom from right ventricle to pulmonary artery reintervention. (E) Freedom from truncal valve reintervention.

Results

Baseline characteristics

Table 1 describes the baseline characteristics of the patients. Among 42 patients who underwent complete repair of TA, 24 patients (57.1%) were neonates, and 3 patients (7.1%) were born prematurely. The median age and weight at operation were 26 days and 3.5 kg, respectively. According to the Collett and Edwards classification, 27 cases (64.3%) were categorized as type I, 11 (26.2%) as type II, and 4 (9.5%) as type III. The morphology of the TrV leaflets was tricuspid in 24 patients (57.1%), quadricuspid in 11 (26.2%), and bicuspid in 7 patients (16.7%). Preoperatively, significant TrV regurgitation was observed in 7 patients (16.7%) and significant TrV stenosis in 11 patients (26.2%). Arch obstruction was also present in 6 patients (14.3%) and 1 patient (2.4%) had a single coronary artery. Chromosomal anomalies were present in 8 patients (19.0%).

Perioperative data

Table 2 presents operative details. One-stage complete repair was performed in 30 patients (71.4%). In 12 patients (28.6%), initial palliation, including truncal separation with RV-PA shunt (n=7), pulmonary artery banding (n=4) and systemic to pulmonary artery shunt (n=1), was performed. To establish the RV-PA connection, a conduit was used in 37 patients (88.1%) and a direct connection (réparation à l'étage ventriculaire [REV] procedure) was per-

Table 4. Factors associated with overall mortality (n=10)

Variable	Hazard ratio (95% confidence interval)	p-value
Univariable Cox regression analysis for overall mortality		
Sex		
Male	Reference	
Female	0.40 (0.08–1.89)	0.248
Prematurity	4.32 (0.89–20.99)	0.069
Age at operation (day)	1.00 (1.00–1.00)	0.065
Low weight at operation (<3.0 kg)	2.66 (0.75–9.43)	0.131
Truncus arteriosus type		
I	Reference	
II	0.87 (0.18–4.33)	0.867
III	2.31 (0.47–11.45)	0.305
No. of TrV cusps		
3	Reference	
2 or 4	2.13 (0.60–7.57)	0.240
Significant TrV dysfunction[a]	3.36 (0.95–11.96)	0.061
Arch obstruction	1.63 (0.34–7.76)	0.535
Chromosomal or noncardiac anomaly	0.89 (0.19–4.26)	0.887
Surgical strategy		
One-stage	Reference	
Staged	0.25 (0.03–1.95)	0.185
Type of right ventricle to pulmonary artery connection		
Conduit	Reference	
Réparation à l'étage ventriculaire	3.79 (0.97–14.80)	0.055
Concomitant TrV surgery	0.60 (0.08–4.76)	0.630
CPB time	1.01 (1.00–1.02)	0.043
Aortic cross-clamp time	1.00 (0.99–1.02)	0.700
Era		
2011 or later	Reference	
Before 2011	4.80 (1.00–22.90)	0.049
Multivariable Cox regression analysis for overall mortality		
Significant TrV dysfunction	6.42 (1.56–26.34)	0.010
CPB time	1.01 (1.00–1.02)	0.018
Era (before 2011)	14.91 (2.39–92.86)	0.004

TrV, truncal valve; CPB, cardiopulmonary bypass.
[a]Significant TrV dysfunction was defined as truncal stenosis or regurgitation greater than mild.

formed in 5 patients (11.9%). The PTFE membrane valved conduit and Contegra (Medtronic Inc., Minneapolis, MN, USA) were the most used RV-PA conduits. The median diameter of the conduit was 12 mm. Concomitant TrV surgery was required in 7 patients (16.7%). The median duration of postoperative mechanical ventilation, intensive care unit stay, and postoperative hospital stay were 6 days, 10 days, and 21 days, respectively. Chylothorax was the most common postoperative complication (n=6, 14.3%).

Survival

There were 8 early deaths (19.0%). In the univariable analysis, the earlier era was associated with early mortality (p=0.031) (Table 3). The overall survival rate was 78.4% at 1 year and 73.8% at 10 years, with a median follow-up duration of 5.8 years (Fig. 1A). In the multivariable analysis, the earlier era (p=0.004), significant TrV dysfunction (p=0.010) and longer cardiopulmonary bypass time (p=0.018) were identified as risk factors for death or transplantation during follow-up (Table 4). Overall survival might have been better in the recent cohort (p=0.028) (Fig. 1B).

Reinterventions

During follow-up, reintervention was required in 27 patients at a median of 1.7 years after complete repair (Table 5). The freedom from all-cause reintervention rates at 1, 5, and 10 years were 79.4%, 31.9%, and 19.1%, respectively (Fig. 1C). In the multivariable analysis, arch obstruction (p<0.001) and significant TrV dysfunction (p=0.011) were identified as independent risk factors for all-cause reintervention (Table 6).

RV-PA reintervention was required in 22 patients at a median of 2.5 years after complete repair (Table 5). The freedom from RV-PA reintervention rates at 1, 5, and 10 years were 82.4%, 36.4%, and 30.9%, respectively (Fig. 1D). In the multivariable analysis, concomitant arch obstruction was identified as an independent risk factor associated with RV-PA reintervention during follow-up (p=0.001) (Table 7).

TrV reintervention was required in 8 patients at a median of 3.8 years after repair during follow-up (Table 5). The cause of TrV reintervention was TrV regurgitation in 7 patients and TrV stenosis in 1 patient. The freedom from TrV reintervention rates at 1, 5, and 10 years were 97.1%, 84.2%,

Table 5. Details of reinterventions

Variable	Value
Patients with all-cause reinterventions	27/34 (79.4)
Total no. of all-cause reinterventions	47
Interval from total repair to all-cause reinterventions (yr)	1.7 (0.3–3.0)
Patients with RV-PA reinterventions	22/34 (64.7)
Interval from index operation to RV-PA reinterventions (yr)	2.5 (1.3–4.2)
Type of reintervention	
RV-PA conduit change with valved conduit	15/22 (68.2)
RVOTR	3/22 (13.6)
Conduit stent insertion	2/22 (9.1)
RV-PA conduit change with valveless conduit	1/22 (4.5)
PVR	1/22 (4.5)
Patients with PA reinterventions	17/34 (50.0)
Interval from index operation to PA reintervention (yr)	2.0 (0.7–3.7)
Patients with TrV reinterventions	8/42 (19.0)
Total no. of TrV reinterventions	8
Interval from total repair to TrV reinterventions (yr)	3.8 (0.7–12.7)
Type of reintervention	
Replacement	6/8 (75.0)
Repair	2/8 (25.0)
Patients with other-cause reinterventions	2/42 (4.8)
Total no. of other-cause reinterventions	2
LVOTR (subaortic membrane resection)	1/2 (50.0)
Ascending aorta graft interposition	1/2 (50.0)

Values are presented as number (%) or median (interquartile range).
RV-PA, right ventricle to pulmonary artery; RVOTR, right ventricular outflow tract reconstruction; PVR, pulmonary valve replacement; PA, pulmonary artery; TrV, truncal valve; LVTOR, left ventricular outflow tract reconstruction.

Table 6. Factors associated with all-cause reintervention (n=27)

Variable	Hazard ratio (95% confidence interval)	p-value
Univariable Cox regression analysis for all-cause reintervention		
Sex		
Male	Reference	
Female	1.12 (0.50–2.48)	0.781
Prematurity	1.47 (0.34–6.35)	0.605
Age at operation (day)	1.00 (1.00–1.00)	0.173
Low weight at operation (<3.0 kg)	2.35 (0.84–6.57)	0.102
Truncus arteriosus type		
I	Reference	
II	0.78 (0.32–1.89)	0.584
III	0.54 (0.12–2.39)	0.419
No. of TrV cusps		
3	Reference	
2 or 4	2.09 (0.92–4.73)	0.078
Significant TrV dysfunction[a]	2.96 (1.24–7.07)	0.014
Arch obstruction	6.64 (2.26–19.54)	0.001
Coronary artery anomaly	0.99 (0.13–7.42)	0.992
Chromosomal or noncardiac anomaly	0.79 (0.27–2.30)	0.664
Surgical strategy		
One-stage	Reference	
Staged	1.02 (0.46–2.27)	0.953
Type of right ventricle to pulmonary artery connection		
Conduit	Reference	
REV	0.22 (0.03–1.65)	0.141
Type of conduit		
PTFE valveless	Reference	
PTFE valved	2.48 (0.79–7.82)	0.122
Contegra	1.80 (0.66–4.92)	0.249
Homograft	0.57 (0.15–2.21)	0.418
REV	0.24 (0.03–1.94)	0.180
Others	0.46 (0.06–3.88)	0.475
Conduit diameter	0.92 (0.76–1.11)	0.370
Concomitant TrV surgery	1.61 (0.60–4.33)	0.346
Cardiopulmonary bypass time	1.00 (0.99–1.01)	0.847
Aortic cross-clamp time	1.00 (0.99–1.01)	0.712
Multivariable Cox regression analysis for all-cause reintervention		
Arch obstruction	7.18 (2.37–21.71)	<0.001
Significant TrV dysfunction	3.14 (1.29–7.63)	0.011
Low weight at operation (<3.0 kg)	2.64 (0.84–8.26)	0.096
No. of TrV cusps (other than 3)	1.25 (0.50–3.13)	0.636

TrV, truncal valve; REV, réparation à l'étage ventriculaire; PTFE, polytetrafluoroethylene.
[a]Significant TrV dysfunction was defined as truncal stenosis or regurgitation greater than mild.

and 68.6%, respectively (Fig. 1E). In the multivariable analysis, significant TrV dysfunction was identified as an independent risk factor associated with TrV reintervention during follow-up (p=0.002) (Table 8).

Discussion

Despite recent improvements in outcomes after neonatal surgery, the risk of mortality after TA repair is not negligible [6,7,10,11,16]. In previous reports, the early mortality rate was 7%–23% [6,10-12,16]. Significant TrV dysfunction, a larger diameter of the right ventricular outflow tract (RVOT) conduit, neonatal surgery, and low body weight have been identified as risk factors for operative mortality in previous studies [6,7,10]. In our study, the early mortality rate was higher than that of the current series; however,

https://doi.org/10.5090/jcs.22.106

Table 7. Factors associated with RV-PA reintervention (n=22)

Variable	Hazard ratio (95% confidence interval)	p-value
Univariable Cox regression analysis for RV-PA reintervention		
Sex		
Male	Reference	
Female	1.01 (0.42–2.45)	0.984
Prematurity	2.10 (0.48–9.18)	0.326
Age at operation (day)	1.00 (1.00–1.00)	0.527
Weight at operation (kg)	0.97 (0.84–1.13)	0.719
Low weight at operation (<3.0 kg)	1.02 (0.30–3.50)	0.978
Truncus arteriosus type		
I	Reference	
II	0.60 (0.21–1.65)	0.320
III	0.24 (0.03–1.83)	0.167
No. of TrV cusps		
3	Reference	
2 or 4	2.94 (1.19–7.29)	0.020
Significant TrV dysfunction[a]	1.48 (0.56–3.92)	0.426
Arch obstruction	3.13 (1.03–9.52)	0.044
Coronary artery anomaly	1.66 (0.22–12.65)	0.626
Chromosomal or noncardiac anomaly	0.44 (0.10–1.88)	0.264
Surgical strategy		
One-stage	Reference	
Staged	1.29 (0.54–3.09)	0.569
Type of RV-PA connection		
Conduit	Reference	
REV	0.32 (0.04–2.40)	0.268
Type of conduit		
PTFE valveless	Reference	
PTFE valved	1.15 (0.29–4.57)	0.839
Contegra	1.19 (0.39–3.63)	0.763
Homograft	0.75 (0.19–2.93)	0.677
REV	0.31 (0.04–2.52)	0.272
Others	0.58 (0.07–4.94)	0.620
Conduit diameter	0.94 (0.77–1.16)	0.577
Concomitant TrV surgery	0.42 (0.10–1.83)	0.251
Cardiopulmonary bypass time	1.00 (0.99–1.01)	0.945
Aortic cross-clamp time	1.00 (0.98–1.01)	0.429
Multivariable Cox regression analysis for RV-PA reintervention		
Arch obstruction	3.60 (1.15–11.23)	0.027
No. of TrV cusps (2 or 4)	3.18 (1.26–7.98)	0.014
Type of RV-PA connection (REV)	0.33 (0.04–2.53)	0.288

RV-PA, right ventricle to pulmonary artery; TrV, truncal valve; REV, réparation à l'étage ventriculaire; PTFE, polytetrafluoroethylene.
[a]Significant TrV dysfunction was defined as truncal stenosis or regurgitation greater than mild.

given that the earlier era was associated with early mortality, early mortality in the current era was much better (4.5%, 1/22) which was comparable with other recent series [5,9]. Furthermore, later attrition among early survivors rarely occurred; consequently, most mortality occurred in the first postoperative year, which is similar to the findings of previous reports [11,12].

The earlier era of surgery and significant TrV dysfunction were identified as factors associated with overall mortality in our study. Significant TrV dysfunction, if concomitantly repaired, would increase surgical complexity and cardiopulmonary bypass time, and, if left untouched during complete repair, would have a negative effect on the ventricle due to a persistently increased workload. Regardless of the initial mode of TrV dysfunction, 3 patients had significant TrV regurgitation before death. Although the small

Table 8. Factors associated with TrV reintervention (n=8)

Variable	Hazard ratio (95% confidence interval)	p-value
Univariable Cox regression analysis for TrV reintervention		
Sex		
Male	Reference	
Female	3.14 (0.74–13.31)	0.121
Prematurity	4.51 (0.52–39.03)	0.171
Age at operation (day)	1.00 (0.99–1.00)	0.259
Weight at operation (kg)	0.80 (0.56–1.14)	0.216
Low weight at operation (<3.0 kg)	5.30 (1.22–22.99)	0.026
Truncus arteriosus type		
I	Reference	
II	1.23 (0.22–6.75)	0.810
III	2.69 (0.48–14.95)	0.258
No. of TrV cusps		
3	Reference	
2 or 4	1.75 (0.43–7.05)	0.432
Significant TrV dysfunction[a]	14.02 (2.72–72.12)	0.002
Arch obstruction	1.06 (0.13–8.85)	0.954
Coronary artery anomaly	0.05 (0.00–114924.52)	0.682
Chromosomal or noncardiac anomaly	5.28 (1.28–21.69)	0.021
Surgical strategy		
One-stage	Reference	
Staged	0.57 (0.11–2.83)	0.491
Type of right ventricle to pulmonary artery connection		
Conduit	Reference	
Réparation à l'étage ventriculaire	0.04 (0.00–349.54)	0.486
Concomitant TrV surgery	7.08 (1.75–28.70)	0.006
Cardiopulmonary bypass time	1.01 (1.00–1.02)	0.052
Aortic cross-clamp time	1.01 (0.99–1.03)	0.270
Multivariable Cox regression analysis for TrV reintervention		
Significant TrV dysfunction	13.83 (2.52–75.79)	0.002
Low weight at operation (<3.0 kg)	4.86 (0.86–27.56)	0.075
Concomitant TrV surgery	3.45 (0.69–17.12)	0.130

TrV, truncal valve.
[a]Significant TrV dysfunction was defined as truncal stenosis or regurgitation greater than mild.

number of events might preclude definitive conclusions in our study, it could be inferred that timely TrV intervention for regurgitant TrV before the progression of LV dysfunction and avoiding over-correction of TrV for relieving the obstruction at the cost of aggravating regurgitation might be critical in order to reduce the risk of death. From a technical standpoint for TrV repair, since several surgical techniques for TrV repair have been introduced, individual surgeons could choose a surgical technique appropriate for the valve pathology. In addition, if valve repair seems not feasible, it could be helpful to lower the threshold for valve replacement because the TrV annulus is usually large enough or easily enlarged anteriorly for placing a commercial valve prosthesis.

The association of the earlier era with overall mortality reflects recent improvements in overall survival (Fig. 1B). When comparing baseline and operative characteristics between the 2 eras (Table 9), repair at our center is now performed earlier, exclusively using a conduit on the RVOT, and taking a longer time for the intracardiac procedure. From these findings, it could be inferred that recent improvements in survival outcomes after TA repair might be attributable to earlier, simpler, and more precise repair.

With respect to mortality outcomes, a point that should be mentioned regarding our surgical strategy is that we have been open to a staged option for patients having conditions not suitable for long surgery using cardiopulmonary bypass. Although early total repair is known to be a prevalent strategy for TA repair [4], staged repair is performed at some centers with comparable outcomes [8].

Table 9. Comparison of baseline and operative characteristics according to the era

Variable	Earlier era (n=20)	Later era (n=22)	p-value
Age at operation (day)	34 (18–565)	12 (9–196)	0.040
Neonate at operation	10 (50.0)	14 (63.6)	0.533
Prematurity	1 (5)	2 (9.1)	1.000
Weight at operation (kg)	3.65 (3.23–9.93)	3.47 (3.12–5.38)	0.392
Low weight at operation (<3 kg)	4 (20)	5 (22.7)	1.000
Sex			0.121
Male	15 (75)	11 (50.0)	
Female	5 (25)	11 (50.0)	
Truncus arteriosus type			0.514
I	12 (60)	15 (68.2)	
II	5 (25)	6 (27.3)	
III	3 (15)	1 (4.5)	
No. of TrV cusps			1.000
3	11 (55)	13 (59.1)	
2 or 4	9 (45)	9 (40.9)	
Significant TrV dysfunction[a]	5 (25)	9 (40.9)	0.338
Associated arch obstruction	1 (5)	5 (22.7)	0.187
Chromosomal or noncardiac anomalies	3 (15)	7 (31.8)	0.284
Surgical strategy			0.738
One-stage	15 (75)	15 (68.2)	
Staged	5 (25)	7 (31.8)	
Type of right ventricle to pulmonary artery connection			0.018
Conduit	15 (75)	22 (100.0)	
Réparation à l'étage ventriculaire	5 (25)	0	
Concomitant TrV surgery	2 (10)	5 (22.7)	0.414
Cardiopulmonary bypass time (min)	163 (127–236)	202 (150–241)	0.170
Aortic cross-clamp time (min)	75 (53–88)	92 (61–131)	0.048

Values are presented as median (interquartile range) or number (%).
TrV, truncal valve.
[a]Significant TrV dysfunction was defined as truncal stenosis or regurgitation greater than mild.

Staged repair was performed in 12 patients (28.6%) and its survival outcome seemed to be favorable (hazard ratio, 0.25; p=0.185). However, given that 3 patients who underwent initial palliation during the study period died before complete repair, it is difficult to make generalizations about the application of the staged approach.

In our study, reinterventions after TA repair were common, as previously reported [7,12,17]. The majority involved RV-PA or TrV reinterventions. In some centers, autologous tissue has been preferred for establishing RV-PA continuity to reduce the risk of RV-PA reintervention [5]; however, RV-PA reintervention might not be inevitable, since non-autologous materials such as homografts, jugular vein valved conduits, prosthetic conduits with bioprosthetic valves, and prosthetic valved conduits have been the predominant choices of material for establishing RV-PA continuity at most centers. Although a pulmonary homograft might have the greatest durability, followed by an aortic homograft, these methods were not easily available in Korea during the study period; moreover, there is a lack of conclusive evidence regarding the superiority of homografts in longevity compared to the other types of conduits [4]. In our study, the 5 patients who underwent the REV procedure did not show superior outcomes regarding freedom from RVOT reintervention. In addition, neither conduit material was superior to the other conduit material in our study. While a smaller diameter of the RV-PA conduit was identified as a risk factor for RV-PA reintervention in previous studies [6,7,17], conduit diameter was not associated with RV-PA reintervention in this study. Interestingly, however, concomitant arch obstruction and a number of TrV cusps other than 3 were identified as independent risk factors for RV-PA reintervention. One possible explanation for the association of arch obstruction with RV-PA reintervention is that an unfavorable course from the right ventricle to the pulmonary artery and a tight space at the distal end of the RV-PA conduit, which were more likely to be present in cases with combined arch obstruction, might be

related to earlier failure of the RV-PA conduit requiring reintervention. Even though the association of the number of TrV cusps with RV-PA reintervention is difficult to explain, since TrV reintervention might be more frequently required in patients who had other than tricuspid TrV, it could be reasonably presumed that patients who had other than tricuspid TrV needed more frequent RV-PA reinterventions due to a lower threshold for RVPA reintervention.

As demonstrated in previous studies, we found that the risk of TrV reintervention was associated with the presence of significant TrV dysfunction at repair [5,7]. Concomitant TrV repair was significantly associated with TrV reintervention in the univariable analysis; however, since it was eliminated in multivariable analysis, it could be speculated that initial TrV dysfunction might play a critical role in the fate of TrV function and that the repair threshold for TrV dysfunction might have been quite high in this study.

This study was limited by the inherent nature of a retrospective single-center study design. Since the small size of the study cohort and the small number of events of interest could weaken the statistical power of all the analyses, readers should be careful in interpreting the results of this study. The changes in surgical skills and perioperative management throughout the 30-year study period might have affected the outcomes despite a sophisticated statistical analysis.

In conclusion, survival outcomes after TA repair improved substantially over the 30-year study period, although reinterventions were required in a significant number of patients during follow-up. Most reinterventions were related to the RV-PA connection or TrV.

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Conflict of interest

No potential conflict of interest relevant to this article was reported.

Funding

This research did not receive any specific grant from funding agencies in the public, commercial, or not-for-profit sectors.

References

1. Hoffman JI, Kaplan S. *The incidence of congenital heart disease*. J Am Coll Cardiol 2002;39:1890-900. https://doi.org/10.1016/s0735-1097(02)01886-7
2. Reller MD, Strickland MJ, Riehle-Colarusso T, Mahle WT, Correa A. *Prevalence of congenital heart defects in metropolitan Atlanta*, 1998-2005. J Pediatr 2008;153:807-13. https://doi.org/10.1016/j.jpeds.2008.05.059
3. Calder L, Van Praagh R, Van Praagh S, et al. *Truncus arteriosus communis: clinical, angiocardiographic, and pathologic findings in 100 patients*. Am Heart J 1976;92:23-38. https://doi.org/10.1016/s0002-8703(76)80400-0
4. Hazekamp MG, Barron DJ, Dangel J, et al. *Consensus document on optimal management of patients with common arterial trunk*. Eur J Cardiothorac Surg 2021;60:7-33. https://doi.org/10.1093/ejcts/ezaa423
5. Derridj N, Villemain O, Khoshnood B, et al. *Outcomes after common arterial trunk repair: impact of the surgical technique*. J Thorac Cardiovasc Surg 2021;162:1205-14. https://doi.org/10.1016/j.jtcvs.2020.10.147
6. Naimo PS, Bell D, Fricke TA, et al. *Truncus arteriosus repair: a 40-year multicenter perspective*. J Thorac Cardiovasc Surg 2021;161:P230-40. https://doi.org/10.1016/j.jtcvs.2020.04.149
7. Guariento A, Doulamis IP, Staffa SJ, et al. *Long-term outcomes of truncus arteriosus repair: a modulated renewal competing risks analysis*. J Thorac Cardiovasc Surg 2022;163:224-36. https://doi.org/10.1016/j.jtcvs.2021.01.136
8. Ota N, Tachimori H, Hirata Y, et al. *Contemporary patterns of the management of truncus arteriosus (primary versus staged repair): outcomes from the Japanese National Cardiovascular Database*. Eur J Cardiothorac Surg 2022;61:787-94. https://doi.org/10.1093/ejcts/ezab348
9. Hrfi A, Ismail M, Mohammed MH, et al. *Outcome of truncus arteriosus repair: 20 years of single-center experience comparing early versus late surgical repair*. Cardiol Young 2022;32:1289-95. https://doi.org/10.1017/S104795112100408X
10. Mastropietro CW, Amula V, Sassalos P, et al. *Characteristics and operative outcomes for children undergoing repair of truncus arteriosus:*

a contemporary multicenter analysis. J Thorac Cardiovasc Surg 2019;157:2386-98. https://doi.org/10.1016/j.jtcvs.2018.12.115

11. Naimo PS, Fricke TA, Yong MS, et al. *Outcomes of truncus arteriosus repair in children: 35 years of experience from a single institution*. Semin Thorac Cardiovasc Surg 2016;28:500-11. https://doi.org/10.1053/j.semtcvs.2015.08.009
12. Tlaskal T, Chaloupecky V, Hucin B, et al. *Long-term results after correction of persistent truncus arteriosus in 83 patients*. Eur J Cardiothorac Surg 2010;37:1278-84. https://doi.org/10.1016/j.ejcts.2009.12.022
13. Collett RW, Edwards JE. *Persistent truncus arteriosus; a classification according to anatomic types*. Surg Clin North Am 1949;29:1245-70. https://doi.org/10.1016/s0039-6109(16)32803-1
14. Baumgartner H, Hung J, Bermejo J, et al. *Echocardiographic assessment of valve stenosis: EAE/ASE recommendations for clinical practice*. J Am Soc Echocardiogr 2009;22:1-23. https://doi.org/10.1016/j.echo.2008.11.029
15. Zoghbi WA, Enriquez-Sarano M, Foster E, et al. *Recommendations for evaluation of the severity of native valvular regurgitation with two-dimensional and Doppler echocardiography*. J Am Soc Echocardiogr 2003;16:777-802. https://doi.org/10.1016/S0894-7317(03)00335-3
16. Russell HM, Pasquali SK, Jacobs JP, et al. *Outcomes of repair of common arterial trunk with truncal valve surgery: a review of the society of thoracic surgeons congenital heart surgery database*. Ann Thorac Surg 2012;93:164-9. https://doi.org/10.1016/j.athoracsur.2011.04.121
17. Bonilla-Ramirez C, Ibarra C, Binsalamah ZM, et al. *Right ventricle to pulmonary artery conduit size is associated with conduit and pulmonary artery reinterventions after truncus arteriosus repair*. Semin Thorac Cardiovasc Surg 2022;34:1003-9. https://doi.org/10.1053/j.semtcvs.2021.05.013