A modified approach in the repair of type I and II truncus arteriosus to promote branch pulmonary arterial growth and limit early reoperation

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ABSTRACT

Objective: Maintaining adequate branch pulmonary arterial growth is critical in preventing early (<3 years) right ventricular outflow tract reoperation after the repair of truncus arteriosus. We hypothesized that a modified truncus arteriosus repair keeping the branch pulmonary arteries in situ would promote branch pulmonary arterial growth and limit early right ventricular outflow tract reoperation.

Methods: For infants requiring repair for type I and II truncus arteriosus, the truncal root was septated through a hockey stick incision keeping the branch pulmonary arteries in situ, the ventricular septal defect was closed, and a short aortic homograft was used to reconstruct the right ventricular outflow tract. Echocardiograms measured preoperative and follow-up branch pulmonary artery diameter.

Results: Between 1998 and 2020, 41 infants were repaired using the modified approach (type I, 28; type II, 13). With a median follow-up of 11.6 (interquartile range, 3.1-15.5) years, there was no significant change between preoperative left pulmonary artery and right pulmonary artery Z-scores and their corresponding follow-up measurement (left pulmonary artery: 0.97, interquartile range, 0.6-1.6 vs left pulmonary artery: 1.4, interquartile range, −0.3 to 1.9) (right pulmonary artery: 0.6, interquartile range, −0.4 to 1.7 vs right pulmonary artery: 0.3 interquartile range, 0.5-0.9). Only 7.3% (n = 2) of follow-up right pulmonary artery Z-scores were less than 2.5 Z-scores below preoperative measurements. Four children (9.8%) required early right ventricular outflow tract reoperation. On multivariable analysis, larger conduit Z-scores were associated with greater time to right ventricular outflow tract reoperation (hazard ratio, 0.55, confidence interval, 0.307-0.984; P = .043).

Conclusions: Maintaining the branch pulmonary arteries in situ at initial truncus arteriosus repair allows for branch pulmonary arterial growth, limiting early right ventricular outflow tract reoperation. (JTCVS Techniques 2022;16:196-211)

Video clip is available online.
likely contribute to the timing of reoperation, potential distortion of the branch PAs at the time of initial repair is often overlooked. Traditionally, the branch PAs are excised from the truncal root and the corresponding defect used as the pulmonary bifurcation. Unfortunately, despite aggressive mobilization, manipulation of the branch PAs during the initial TA repair can result in multilevel obstruction and hypoplasia, which can increase the risk for conduit failure and early RVOT reoperation.5,10 Since 1998, we have used a modified approach in the repair of TA where the branch PAs are kept in situ and a short aortic homograft used to establish right ventricular to PA continuity.11-13 Using this approach exclusively, we have demonstrated excellent freedom from RVOT reoperation as well a limited need for surgical pulmonary arterioplasty during short- and intermediate-term follow-up. However, long-term outcomes, potential for branch PA growth, and risks for early reoperation using this modified approach have been left unexplored. We hypothesized that using a modified approach to repair TA during early infancy would promote interval growth in the branch PAs, limiting early RVOT reoperation.

MATERIALS AND METHODS
After Institutional Review Board approval by the University of Rochester Medical Center (RSRB No.: 00006298, approved on 6/4/2021) and waiver of consent, all infants with type I or II TA14 repaired between 1998 and 2020 using our modified approach were reviewed.

Operative Technique
A detailed description of the modified approach in the repair of TA has been published,11 along with a prior video demonstrating the steps of the technique.13 After aortic and venous cannulation and placement of a left ventricular vent, snares are placed around the branch PAs, cardiopulmonary bypass instituted, and antegrade cardioplegia used to arrest the heart in a standard fashion. A hockey-stick incision on the truncal root curving onto the left pulmonary artery (LPA) is used to evaluate the defect among the aorta PA, the truncal valve, the coronary arteries, and the PAs. If the truncal valve requires repair or replacement, it can be performed using this incision. A 0.4-mm polytetrafluoroethylene (Bard) patch is used to septate the truncal root from the PAs. The septation patch for type I TA is often circular, due to the distance between the branch PAs and the defect.

FIGURE 1. Representative intraoperative photographs for the (A) incision on the pulmonary trunk and right ventriculotomy, (B) aortic homograft, and (C and D) trimming and preparing the aortic homograft demonstrating that the anterior leaflet of the mitral valve is used as a hood for the proximal anastomosis. (E) Completion of the proximal anastomosis, (F and G) the distal anastomosis demonstrating that tissue just distal to the aortic homograft valve annulus is sewn to the PA bifurcation, and (H) completed TA repair.

Abbreviations and Acronyms
CI = confidence interval
CMR = cardiac magnetic resonance
IQR = interquartile range
LPA = left pulmonary artery
PA = pulmonary artery
RPA = right pulmonary artery
RVOT = right ventricular outflow tract
TA = truncus arteriosus
For infants with type II TA, the septation patch is often more oblong, because the right pulmonary artery (RPA) and LPA exit the truncus separately.

After septation of the truncal root, the previous hockey stick incision becomes the site of the distal anastomosis for the aortic homograft. Through a right ventriculotomy, the ventricular septal defect is patchsed using 0.4-mm polytetrafluoroethylene (Bard). An aortic valved homograft is sized to the PA bifurcation and selected on the basis of availability. The proximal anastomosis is completed first, using the anterior leaflet of the mitral valve as a hood sutured to the RVOT. The majority of the posterior facing sinus of the homograft is excised, along with lateral and medial tissue so that only the aortic homograft valve annulus remains circumferential and is sewn to the PA bifurcation (Figure 1, A-H).

Right Ventricular Outflow Tract Reoperation
RVOT reoperation was performed when (1) the peak right ventricular outflow gradient was greater than 75 mm Hg as measured by echocardiogram, (2) cardiac catheterization data demonstrated a RVOT gradient of greater than three-fourths the systemic arterial pressure, or (3) in the presence of RVOT insufficiency with a dilated right ventricle and corresponding right ventricular end-diastolic index greater than 200 mL/m². At the time of reoperation, after dissection of the mediastinal contents, an aortic cannula was placed within the aortic arch, and bicaval or single venous drainage was used to initiate cardiopulmonary bypass. The previously placed aortic homograft was identified and excised at the level of the branch PAs and right ventricular suture lines. The tissue adjacent to the suture lines was freshened, and inspection of the branch PAs was performed. If there was any evidence of obstruction or if the distal anastomotic site was insufficient in size, the previously placed septation patch was identified, and the RPA cut off the patch, leaving the patch intact on the truncal root. The use of cardioplegia to arrest the heart has not been necessary to disconnect the RPA from the truncal root. Once the distal anastomosis site was established, a PA valved xenograft conduit was brought into the field. The distal anastomosis and subsequent proximal anastomoses were performed, the patient was weaned from cardiopulmonary bypass, and the chest was closed. A surgical video of conduit exchange after the modified approach has been previously published and offers further visual assistance.

Study Measures
Serial echocardiographic measurements quantified the peak RVOT gradient and the degree of pulmonary insufficiency (trivial, mild, moderate, and severe) over time. To evaluate for changes in somatic growth in relation to the fixed size of the aortic homograft, the pulmonary valve Z-score was calculated on the basis of follow-up body surface area. The initial postoperative echocardiogram and Z-scores were defined as time zero. Spaghetti plots were constructed for the pulmonary valve Z-score, peak RVOT gradient, and degree of pulmonary insufficiency. A linear regression with 95% confidence intervals (CIs) was constructed to demonstrate the effect over time.

The LPA and RPA were measured from the mid-section of the vessel preoperatively and either at the time of reoperation or most recent follow-up. Corresponding Z-scores were calculated on the basis of body surface area, and spaghetti plots were constructed to illustrate the change in Z-score over time. The Nakata index was calculated at the same time points as previously described.

When available, cardiac catheterization data measured mean pressures within the RVOT distal to the valve (main PA) and branch PAs. Gradients between the main and branch PAs and between each branch PA, and the number of pressure gradients greater than 10 mm Hg were calculated.

Outcome Measures
The primary outcome was to assess the growth in the LPA and RPA Z-scores from preoperative to follow-up measurements and identify variables associated with earlier RVOT reoperation within 3 years of initial TA repair. Early RVOT reoperation was defined as within 3 years, given our previous experience that conduit replacement before 3 years of age results in several reoperations before adulthood. Secondary outcomes included freedom from surgical branch pulmonary arterioplasty and freedom from RVOT reoperation.

Statistics
Descriptive statistics were used to describe the study measures, such as mean with standard deviation, median with interquartile range (IQR), and frequency with percentage. Continuous variables were compared between

TABLE 1. Perioperative demographics

| Preoperative details | Truncus (n = 41) |
|----------------------|-----------------|
| Age (d)              | 16.0 (9-15)     |
| Male gender          | 48.7% (20)      |
| Weight (kg)          | 3.2 (2.7-3.6)   |
| Prematurity < 37 wk  | 7.3% (3)        |
| TA type              |                 |
| Type I               | 70.7% (29)      |
| Type II              | 29.3% (12)      |
| DiGeorge syndrome    | 24.4% (10)      |
| RAA                  | 24.4% (10)      |
| IAA                  | 19.5% (8)       |
| Coronary abnormality | 17.1% (7)       |
| Truncal valve        |                 |
| Bicuspid             | 4.9% (2)        |
| Tricuspid            | 46.3% (19)      |
| Quadracuspid         | 46.3% (19)      |
| Pentacuspid          | 7.1% (1)        |
| More than mild TV stenosis | 21.9% (9) |
| More than mild TV insufficiency | 7.1% (3) |
| Operative details    |                 |
| Cardiopulmonary bypass time (min) | 181.5 (164.5-200.5) |
| Aortic crossclamp time (min) | 129.0 (117.5-141.0) |
| Conduit size (mm)    | 11.0 (10.0-12.0) |
| Conduit Z-score       | 2.7 (1.9-3.1)   |
| Concomitant procedure |                 |
| IAA repair           | 19.5% (8)       |
| Coronary artery unroofing | 2.4% (1) |
| TV repair            | 2.4% (1)        |
| TV replacement        | 2.4% (1)        |
| Postoperative        |                 |
| Delayed sternal closure | 14.6% (6) |
| Duration mechanical ventilation (d) | 4 (3.0-7.0) |
| Mechanical ventilation > 7 d | 12.2% (5) |
| ECMO                 | 0               |
| Pulmonary HTN crisis  | 7.3% (3)        |
| Sepsis               | 4.9% (2)        |
| Mortality            | 2.4% (1)        |
| Hospital LOS (d)     | 15.0 (10.3-27.8) |

TA, Truncus arteriosus; RAA, right aortic arch; IAA, interrupted aortic arch; TV, truncal valve; ECMO, extracorporeal membrane oxygenation; HTN, hypertension; LOS, length of stay.
study groups using a Mann–Whitney or 2-tailed Student t test where appropriate, and categorical variables were compared using the Fisher exact test. Freedom from RVOT reoperation and surgical branch pulmonary arterioplasty was assessed using Kaplan–Meier curves. Linear regression was used to analyze the change in pulmonary valve Z-score over time, peak RVOT gradient (mm Hg), and degree of pulmonary insufficiency. Two-way analysis of variance examined differences between the LPA and RPA dimensions from preoperative and follow-up measurements. Multivariable Cox proportional hazards regression analysis was performed to evaluate the association between identified preoperative and intraoperative variables previously associated with an earlier time to RVOT reoperation, while accounting for the competing risk of mortality. The variables in the model included age at repair, conduit and branch PA size, and the type of TA.

All statistics were completed using SAS, SPSS 21, or GraphPad Prism. A power calculation was not performed because this was a follow-up of infants previously repaired.

RESULTS

A modified repair was performed in 41 sequential infants with type I and II TA. Baseline demographics demonstrated that the majority of children had type I TA, and a significant percentage of those children had associated comorbidities such as DiGeorge syndrome, interrupted aortic arch, and coronary abnormalities (Table 1). The median-sized aortic homograft used to establish right ventricular to PA continuity was 11 mm. There was 1 operative mortality. A 2.2-kg, former 32-week gestation neonate who was severely dysmorphic and had multiple noncardiac anomalies died of fungal sepsis on postoperative day 32. Genetic testing was not performed. Follow-up was obtained in 100% of patients within 2 years or at the time of mortality at a median of 11.6 (IQR, 3.1-15.5) years. The follow-up for imaging studies was 7.3 (IQR, 3.4-12.4) years. Overall survival was 85.6% at 15 years (Figure E1). There were no mortalities associated with subsequent reintervention.

Branch Pulmonary Arteries

Figure 2, A and B illustrate the change in branch PA Z-scores over time. Preoperatively, the LPA and RPA (LPA: 5.0 [IQR, 4.0-6.0 mm] vs RPA: 5.1 [IQR, 4.3-6.3 mm] P = .8), and corresponding Z-scores (LPA: 0.97 IQR, 0.6-1.6 vs RPA: 0.6 IQR, –0.4 to 1.7) were similar in size (Figure 2, C). Follow-up dimensions of the branch
PAs more than doubled and remained similar (LPA: 12.0 [IQR, 8.2-15.5 mm] vs RPA: 12.0 [IQR, 8.0-15.0 mm]; \( P = .9 \)). Although LPA Z-scores were significantly greater than RPA Z-scores at follow-up, there was no significant change between preoperative and follow-up measurements (LPA 1.4 [IQR, –0.3 to 1.9] vs RPA 0.3 [IQR, 0.5-0.9]; \( P = .23 \)).

Likewise, the percentage of negative Z-scores did not significantly change during the study period (Figure 2, D). Only 14% (6) of LPA and 21% (9) of RPA Z-scores decreased by more than 1.0 between preoperative and follow-up measurements. Z-scores less than –2.5 at follow-up were observed in 2 RPA measurements and no LPA measurements. There were no statistically significant differences in the Nakata index over the study period (Figure E2). Cardiac magnetic resonance (CMR) measurements of the branch PA flow are provided in Table E1.

The spaghetti plots for the pulmonary valve Z-scores based on the size of the aortic homograft, peak RVOT gradient, and degree of pulmonary insufficiency are shown in Figure E3, A-C. The median pulmonary valve Z-score decreased over time and dropped below zero by 2 years postoperatively (Figure 3, A). Likewise, median peak RVOT gradients increased over time and plateaued along with the degree of insufficiency at approximately 2 years, often leaving children with moderate mixed prosthetic valvar disease (Figure 3, B and C).

Catheter-based reinterventions on the homograft were performed in 8 children, and freedom from catheter-based reintervention on the homograft was 67% at 10 years. Four children required a catheter-based reintervention on the branch PAs, with a resulting freedom from catheter reintervention on the branch PAs of 87% at 10 years (Figure 4, A and B). Four patients required early RVOT reoperation between 0.5 and 2.7 years (Table E2). The primary indication for reoperation was idiopathic degeneration of the aortic valved homograft (n = 3) or endocarditis (n = 1). Demographics of these patients in comparison with those who did not require early RVOT reoperation are provided in Tables E3, E4, and E5. Children requiring early RVOT reoperation had significantly smaller preoperative LPA Z-scores and smaller aortic homograft Z-scores.

![Figure 3](https://example.com/fig3.png)

**FIGURE 3.** A, Regression and 95% CIs (blue) for the pulmonary valve z-score during follow-up after modified truncus repair. B, Regression and 95% CIs (blue) for peak echocardiographic gradients during follow-up after modified truncus repair. C, Regression and 95% CIs (blue) for the degree of pulmonary insufficiency during follow-up after modified truncus repair. RVOT, Right ventricular outflow tract.
Twenty-five children (60.9%) required RVOT reoperation during the follow-up period (Table 2). Demographics at the time of reoperation are provided in Table 3. At the time of reoperation, 36.0% (9/25) of children required excision of the branch PAs from the truncal root (Video Abstract). For the majority of children (17/25, 68%), the conduit size used at reoperation was greater than or equal to 20 mm. Freedom from RVOT reoperation was 71.2%, 48.7%, and 9.5% at 5, 10, and 15 years, respectively (Figure 4, C). There were no isolated surgical pulmonary arterioplasties without concomitant conduit replacement. Freedom from surgical pulmonary arterioplasty was 93.5%, 88.6%, and 83.7% at 5, 10, and 15 years, respectively (Figure 4, D). Subsequent freedom from a secondary RVOT reintervention after conduit replacement was 75.4% at 10 years (Figure E4). Multivariable analysis to examine the risk factors associated with time to reoperation demonstrated that only larger conduit Z-scores at the time of repair were significantly protective (Table 3 and Figure 5).

**DISCUSSION**

Maintaining the branch PA architecture at the time of TA repair has been reported for approximately 40 years. First described as a case study in 1975, and then as a large case series by Barbero-Marcial and Tanamati, these early reports illustrated the benefits of keeping the branch PAs in situ to prevent multilevel stenosis and hypoplasia. We believe that excision of the branch PAs from the truncal root at the time of TA repair increases the risk for PA distortion that can develop into multilevel obstruction and minimize growth. Similar to our experience, other groups have shown that at baseline children born with TA have normal or greater than normal RPA and LPA Z-scores. This suggests that the development of branch PA hypoplasia is not intrinsic to TA, but rather a result of the repair. By using our modified approach, the median LPA and RPA Z-scores changed by +0.4 and –0.2, respectively, over a follow-up of 10 years. This negligible change illustrates continued branch PA growth and may contribute to a longer freedom from RVOT reoperation.

**FIGURE 4.** Kaplan–Meier curves comparing the freedom from (A) homograft catheter-based intervention, (B) PA catheter-based intervention, (C) RVOT reoperation (Re-Op), and (D) surgical branch pulmonary arterioplasty. For all curves, the shaded blue area represents the 95% CIs, and the subjects at risk placed above the X-axis. RVOT, Right ventricular outflow tract.
TABLE 2. Follow-up at reoperation

| Variable                              | N = 25
|---------------------------------------|--------
| Age (y)                               | 7.3 (3.4-12.4) |
| Weight (kg)                           | 23.0 (14.7-38.5) |
| TV disease                            |        |
| More than mild TV stenosis            | 8.0% (2) |
| More than mild TV insufficiency       | 12.0% (3) |
| RVOT gradient (mm Hg)                 | 62.0 (53.0-71.3) |
| RVOT insufficiency                    |        |
| Mild                                  | 16.0% (4) |
| Moderate                              | 68.0% (17) |
| Severe                                | 16.0% (4) |
| Cardiac catheterization (n = 25)      |        |
| RV (mm Hg)                            | 60.0 (51.0-84.0) |
| MPA (mm Hg)                           | 18.0 (16.0-22.0) |
| LPA (mm Hg)                           | 16.0 (14.5-18.0) |
| RPA (mm Hg)                           | 15.0 (13.0-19.0) |
| RPA/MPA gradient (mm Hg)              | 2.0 (0.0-4.0) |
| RPA/MPA gradient > 10 mm Hg           | 8.0% (2) |
| LPA/MPA gradient (mm Hg)              | 1 (–1 to 3.5) |
| RPA/LPA gradient (mm Hg)              | 0 (0-2.5) |
| RPA/LPA gradient > 10 mm Hg           | 4.0% (1) |
| Reoperative details                  |        |
| RV-PA conduit type                    |        |
| CE valved conduit                     | 40.0% (10) |
| Hancock valved conduit                | 60.0% (15) |
| RV-PA conduit size (mm)               | 22 (18-23) |
| RV-PA conduit Z-score                 | 0.9 (0.5-1.8) |
| Truncal valve repair                  | 4.0% (1) |
| Truncal valve replacement             | 8.0% (2) |
| Branch PA excision                    | 36.0% (9) |
| Branch PA patch                        | 16.0% (4) |

TV: Truncal valve; RVOT, right ventricular outflow tract; RV, right ventricular; MPA, main pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; RV-PA, right ventricular to pulmonary artery.

TABLE 3. Cox regression analysis for time-dependent factors associated with right ventricular outflow tract reoperation

| Variable                              | HR (95% CI) | P value |
|---------------------------------------|-------------|---------|
| Age at repair                         | 1.006 (0.993-1.019) | .369    |
| Type I TA                             | 1.510 (0.690-3.303) | .502    |
| RV-PA conduit Z-score                 | 0.550 (0.307-0.984) | .043    |
| LPA Z-score                           | 0.77 (0.489-1.21) | .770    |
| RPA Z-score                           | 0.947 (0.662-1.355) | .947    |

HR, Hazard ratio; CI, confidence interval; TA, truncus arteriosus; RV-PA, right ventricular to pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery.

reports of branch PA reintervention within the first year after TA repair, the timing of branch PA reoperation using the modified approach is longer (2.8-10.6 years). This further validates that the mechanism behind branch PA stenosis after TA repair with the modified approach is different when the branch PAs are excised and is well tolerated.

Approximately one-third of children required excision of the branch PAs at the time of reoperation to establish a larger orifice for the PA bifurcation. Although excision of the branch PAs at any time point may increase the risk of branch PA distortion, our preference is to excise the branch PAs at reoperation. At reoperation, the branch PAs have often doubled or tripled in size and the risk of distortion from PA excision and subsequent hypoplasia remain minimal.

Freedom from RVOT reoperation after the repair of TA is commonly reported between 30% and 55% at 5 years. Using our modified approach, our group and others have consistently demonstrated a freedom from RVOT reoperation approaching 50% or better at 10 years. Although conduit failure after TA repair is inevitable, by using our modified approach, most children develop both mild to moderate RVOT stenosis and insufficiency by 2 years of age. We believe that the balance of both stenosis and insufficiency, in the absence of branch PA stenosis, extends the freedom of RVOT reoperation. Rapid somatic growth occurs in the first 2 years after repair, as evidenced by the steepest decline in the pulmonary valve Z-score. However, the burden of RVOT stenosis remains limited when the aortic homograft valve annulus remains the only circumferential portion of the conduit, and the remainder of the RVOT continues to grow (posterior RVOT and the PA bifurcation). Use of a pulmonary homograft may yield similar results; however, the construction of a pericardial hood may be necessary to complete the proximal anastomosis.

Limiting initial RVOT reoperation is essential to prevent the second and third reoperations before adulthood. On the basis of univariate statistics, children requiring early reoperation had smaller aortic homografts and smaller LPA Z-scores at the time of TA repair. Only larger aortic...
homograft Z-scores were protective in preventing early reoperation on multivariable analysis. Although larger homografts were protective, at the time of repair the homograft was sized to the PA bifurcation. Therefore, the protection afforded from a larger aortic homograft may be reflective of larger branch PAs.

Multiple early reinterventions and reoperations are associated with increased morbidity and mortality for each subsequent procedure.1,19,20 By extending the time to reoperation and allowing the branch PAs to grow, the placement of a larger-sized conduit (≥20 mm) at the time of the first reoperation allows for future catheter-based valve reintervention.21,22 Children repaired using the modified approach, based on conduit longevity and subsequent catheter reintervention, may not require a third operation until their fifth or sixth decade.

Study Limitations

This study has several limitations. This is a single surgeon series, and there is no comparison group. Data were obtained retrospectively and are limited by that approach. Last, there were a limited number of CMR data available that prevented a more complete assessment of the right ventricular function and branch PA flow. Although there were no differences in branch PA Z-scores between preoperative and follow-up measurements, the variation in the time to follow-up between patients may have influenced the results. Further, the size of the early reoperation cohort was small, and therefore a larger sample size may produce different results.

CONCLUSIONS

Preventing early reoperation after the repair of type I and II TA is critically important. Maintenance of the branch PA architecture using a modified approach for infants prevented multilevel branch PA obstruction and hypoplasia, which we believe helped extend the time to RVOT reoperation. In addition, the selection of a short aortic homograft often leaves the child with mixed RVOT insufficiency and stenosis, which can be well tolerated for many years. This suggests that the use of a modified repair could be considered during the repair of type I and II TA.
Webcast
You can watch a Webcast of this AATS meeting presentation by going to: https://www.aats.org/resources/1500.

Conflict of Interest Statement
The authors reported no conflicts of interest.

The Journal policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have disclosed conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest.

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Key Words: branch pulmonary artery stenosis, reoperation, truncus arteriosus
APPENDIX 1. METHODS

If surgical patch arterioplasty is required, primarily for right PA stenosis, a reoperative sternotomy is performed, the mediastinal adhesions are dissected, and the patient is placed on cardiopulmonary bypass. While the heart remains beating, the previously placed aortic homograft is removed, and the right PA dissected off the posterior surface of the aorta, leaving the septation patch intact on the aorta. The anterior surface of the right PA is opened and angioplastied with PA homograft using 7-0 Prolene. The distal end of the right ventricular to PA conduit is then sewn to the previously established pulmonary bifurcation using 7-0 Prolene, the proximal anastomosis is performed with 5-0 Prolene, and the child is weaned from cardiopulmonary bypass.

CARDIAC MAGNETIC RESONANCE IMAGING

When CMR imaging was performed, right ventricular chamber size, systolic function, and degree of RVOT insufficiency were measured. In addition, if measured, the percent of right PA flow was recorded.

RESULTS

Four children underwent catheter-based reintervention on the branch PAs. Three reinterventions were performed on the proximal right PA without stent implantation. One child required several branch PA catheter reinterventions that include stenting of the mid and distal right PA. This child was a former 10-day-old 2.3-kg neonate with preoperative PA hypertension and chronic lung disease.
**FIGURE E1.** Kaplan–Meier survival curve for 41 children who underwent a modified approach in the repair of TA. The **solid blue line** represents the survival, and **blue shaded area** represents the 95% CIs. The number of subjects at risk are provided above the X-axis.

**FIGURE E2.** Box and whisker plot demonstrating the median (**horizontal line**) and upper (75th percentile) and lower quartiles (25th percentile) of the baseline and follow-up Nakata Index in children after a modified approach to the repair of TA.
FIGURE E3. Spaghetti plots for the (A) pulmonary valve Z-score, (B) peak RVOT gradient, and (C) degree of pulmonary insufficiency from 41 children with TA repaired using a modified approach. RVOT, Right ventricular outflow tract.

FIGURE E4. Freedom from secondary RVOT reintervention in 25 children requiring RVOT reoperation. The solid blue line represents the freedom from secondary freedom from RVOT reintervention, and the blue shaded area represents the 95% CIs. The number of subjects at risk are provided above the X-axis. RVOT, Right ventricular outflow tract.
FIGURE E5. A and B, Representative CMR 3-dimensional reconstruction of a child after TA repair using the modified approach. These images demonstrate that the position of the proximal RPA located leftward off the truncal root and in close association with the septation patch. As the truncal root increases in size, it can push against the lower pressure RPA wall and partially obstruct the RPA orifice (C).
### TABLE E1. Cardiac magnetic resonance imaging data

|                              | After modified truncus repair (n = 7) |
|------------------------------|--------------------------------------|
| Age (y)                      | 11.7 (6.0-15.5)                     |
| RVEDV (mL)                   | 161 (118-188)                       |
| RVEDV index (mL/m²)          | 129.0 (96-202)                      |
| RVEF (%)                     | 54 (45-59)                          |
| PR (%)                       | 29 (24-38)                          |
| RPA flow % (n = 3)           | 40 (25-54)                          |

*RVEDV*, Right ventricular end-diastolic index; *RVEF*, right ventricular ejection fraction; *PR*, pulmonary regurgitation; *RPA*, right pulmonary artery.

### TABLE E2. Characteristics of children requiring early right ventricular outflow tract reoperation

| Age at reoperation | Truncus type | Primary indication          |
|--------------------|--------------|-----------------------------|
| 1.79               | I            | RVOT insufficiency          |
| 2.6                | II/III       | RVOT stenosis/insufficiency |
| 2.7                | II           | RVOT stenosis/insufficiency |
| 0.6                | II           | Homograft endocarditis      |

*RVOT*, Right ventricular outflow tract reoperation.
### TABLE E3. Preoperative demographics of children requiring an early reoperation

|                      | No early reoperation (37) | Early reoperation (4) | P value |
|----------------------|---------------------------|-----------------------|---------|
| Age (d)              | 16 (8.5-33.0)             | 17.5 (15.3-35.5)      | .55     |
| Male gender          | 51.3% (19)                | 25.0% (1)             | .61     |
| Weight (kg)          | 3.2 (2.7-3.6)             | 3.3 (2.7-3.6)         | .81     |
| Prematurity          | 8.1% (3)                  | 0                     | .99     |
| TA type              |                           |                       |         |
| Type I               | 75.6% (28)                | 25.0% (1)             | .07     |
| Type II              | 24.4% (9)                 | 75.0% (3)             | .07     |
| DiGeorge syndrome    | 16.2% (6)                 | 25.0% (1)             | .54     |
| RAA                  | 24.3% (10)                | 0                     | .56     |
| IAA                  | 18.9% (7)                 | 0                     | .99     |
| Coronary abnormality | 14.6% (6)                 | 25.0% (1)             | .54     |
| Branch PAs           |                           |                       |         |
| LPA (mm)             | 5.1 (4.2-6.0)             | 4.4 (4.0-5.0)         | .19     |
| LPA Z-score          | 0.9 (0.2-1.7)             | –0.4 (–1.3 to 0.4)    | .02     |
| RPA (mm)             | 5.3 (4.3-6.7)             | 4.6 (3.9-5.7)         | .30     |
| RPA Z-score          | 0.7 (–0.1 to 1.7)         | –0.4 (–1.6 to 0.8)    | .12     |
| Nakata index         | 219.3 (138.2-284.1)       | 157.6 (121.8-232.4)   | .31     |
| Truncal valve        |                           |                       |         |
| Bicuspid             | 5.4% (2)                  | 0                     | .99     |
| Tricuspid            | 43.2% (16)                | 75.0% (3)             | .32     |
| Quadricuspid         | 48.6% (18)                | 25.0% (1)             | .61     |
| Pentacupid           | 2.7% (1)                  | 0                     | .99     |
| More than mild TV stenosis | 24.3% (9) | 0% (0) | .55 |
| More than mild TV insufficiency | 8.1% (3) | 0 | .99 |

TA, Truncus arteriosus; RAA, right aortic arch; IAA, interrupted aortic arch; PA, pulmonary artery; LPA, left pulmonary artery; RPA, right pulmonary artery; TV, truncal valve.

### TABLE E4. Operative data of children requiring an early reoperation

|                      | No early reoperation (37) | Early reoperation (4) | P value |
|----------------------|---------------------------|-----------------------|---------|
| CPB time (min)       | 189 (164.5-211.0)         | 153.5 (140.0-169.3)   | .01     |
| Aortic crossclamp time (min) | 130.5 (118.8-141.5) | 115.5 (102.5-129.3)   | .11     |
| Conduit size (mm)    | 12.0 (11.0-12.5)          | 10.5 (9.3-11.6)       | .24     |
| Conduit z-value      | 2.8 (2.4-3.1)             | 1.6 (1.0-2.6)         | .05     |
| Concurrent procedure |                           |                       |         |
| IAA repair           | 8                         | 0                     | .56     |
| Coronary artery      | 1                         | 0                     | .99     |
| TV repair            | 0                         | 0                     | .99     |
| TV replacement       | 1                         | 0                     | .99     |
| Postoperative        |                           |                       |         |
| Delayed stern closure| 6                         | (0)                   | .99     |
| Duration mechanical ventilation | 4 (3.0-7.0) | 4.5 (3.0-6.8) | .95     |
| ECMO                 | 0                         | 0                     | .99     |
| Pulmonary HTN crisis | 3                         | 0                     | .99     |
| Sepsis               | 2                         | 0                     | .99     |
| Hospital LOS (d)     | 15.5 (10.3-27.8)          | 12.0 (8.8-28.0)       | .64     |

CPB, Cardiopulmonary bypass; IAA, interrupted aortic arch; TV, truncal valve; ECMO, extracorporeal membrane oxygenation; HTN, hypertension; LOS, length of stay.
| TABLE E5. Follow-up at reoperation |
|----------------------------------|
|                                  | No early reoperation (21) | Early reoperation (4) | P value |
| Age (y)                          | 10.4 (5.8-12.8)           | 2.2 (0.9-2.7)         | <.01    |
| Weight (kg)                      | 30.0 (16.4-45.5)          | 9.8 (7.4-16.9)        | <.01    |
| More than mild TV stenosis       | 9.5% (2)                  | 0                     | .99     |
| More than mild TV insufficiency  | 14.2% (3)                 | 0                     | .99     |
| RVOT gradient (mm Hg)            | 68.0 (53.0-77.0)          | 62.0 (50.3-71.5)      | .53     |
| RVOT insufficiency               |                          |                       |         |
| Mild                             | 19.0% (4)                 | 0                     | .99     |
| Moderate                         | 76.2% (16)                | 25.0% (1)             | .08     |
| Severe                           | 4.8% (1)                  | 75.0% (3)             | <.01    |
| Catheterization                  |                          |                       |         |
| RV (mm Hg)                       | 60.0 (50.0-80.0)          | 87 (66.0-103.5)       | .07     |
| MPA (mm Hg)                      | 29.5 (24.0-33.5)          | 31.0 (24.7-47.0)      | .48     |
| RPA (mm Hg)                      | 21.0 (18.3-24.0)          | 23.0 (20.0-32.8)      | .37     |
| LPA (mm Hg)                      | 26 (18.5-30.5)            | 24.0 (18.3-26.8)      | .65     |
| Operative details                |                          |                       |         |
| Concomitant procedure            |                          |                       |         |
| TV repair                        | 4.8% (1)                  | 0                     | .99     |
| TV replacement                   | 9.5% (2)                  | 0                     | .99     |
| PA plasty                        | 9.5% (2)                  | 50.0% (2)             | .20     |
| Conduit size                     | 22.0 (20.0-25.0)          | 18.0 (15.0-18.0)      | <.01    |
| Conduit Z score                  | 0.8 (0.4-1.5)             | 2.4 (1.3-3.0)         | .02     |

TV, Truncal valve; RVOT, right ventricular outflow tract; RV, right ventricle; MPA, main pulmonary artery; RPA, right pulmonary artery; LPA, left pulmonary artery; PA, pulmonary artery.