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Multicenter Analysis of Early Childhood Outcomes Following Repair of Truncus Arteriosus

Running Head: Truncus Arteriosus Outcomes

[1]Jason R. Buckley, MD; [2]Venu Amula, MD; [3]Peter Sassalos, MD; [4]John M. Costello, MD, MPH; [5]Arthur J. Smerling, MD; [6]lias Iliopoulos, MD; [7]Aimee Jennings, PNP-AC/PC; [8]Christine M. Riley, CPNP-AC; [9]Katherine Cashen, DO; [10]Sukumar Suguna Narasimhulu, MD, MPH; [11]Keshava Murthy Narayana Gowda, MBBS; [12]Adnan M. Bakar, MD; [13]Michael Wilhelm, MD; [14]Aditya Badheka, MD; [15]Elizabeth AS Moser, MS; [16]Christopher W. Mastropietro, MD; and the Collaborative Research in Pediatric Cardiac Intensive Care (CoRe-PCIC) Investigators.

[1] Department of Pediatrics, Medical University of South Carolina Children's Hospital, Charleston, SC

[2] Department of Pediatrics, University of Utah School of Medicine, Primary Children's Hospital, Salt Lake City, UT

[3] Department of Cardiac Surgery, University of Michigan, C.S. Mott Children's Hospital, Ann Arbor, MI

[4] Department of Pediatrics, Ann & Robert H. Lurie Children's Hospital of Chicago, Northwestern University Feinberg School of Medicine

[5] Department of Pediatrics, Columbia University College of Physicians & Surgeons, Morgan Stanley Children's Hospital of New York, New York, NY

[6] Department of Pediatrics, The Heart Institute, Cincinnati Children's Hospital Medical Center, Cincinnati, OH

[7] Department of Pediatrics, Seattle Children's Hospital, Seattle, WA

[8] Department of Pediatrics, Children's National Health System, Washington, DC

[9] Department of Pediatrics, Wayne State University School of Medicine, Children's Hospital of Michigan, Detroit, MI

[10] Department of Pediatrics, University of Central Florida College of Medicine, The Heart Center at Arnold Palmer Hospital for Children, Orlando, FL

[11] Department of Pediatrics, Cleveland Clinic, Cleveland, OH

[12] Department of Pediatrics, Zucker School of Medicine at Hofstra / Northwell, Cohen Children's Medical Center of NY, New Hyde Park, NY

[13] Department of Pediatrics, University of Wisconsin, Madison, WI

[14] Department of Pediatrics, University of Iowa Stead Family Children's Hospital, Iowa City, IA

[15] Department of Biostatistics, Indiana University School of Medicine & Richard M. Fairbanks School of Public Health, Indianapolis, IN

[16] Department of Pediatrics, Indiana University School of Medicine, Riley Hospital for Children, Indianapolis, IN

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Corresponding Author:

Jason R. Buckley, MD

Medical University of South Carolina

601 Children's Hospital

165 Ashley Avenue, MSC 915

Charleston, SC, 29425

Email: buckleyj@musc.edu

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Abstract

Background: Literature describing morbidity and mortality following truncus arteriosus repair is predominated by single-center reports. We created and analyzed a multicenter dataset to identify risk factors for late mortality and right ventricle-to-pulmonary artery (RV-PA) conduit reintervention for this patient population.

Methods: We retrospectively collected data on children who underwent repair of truncus arteriosus without concomitant arch obstruction at 15 centers between 2009 and 2016. Cox regression survival analysis was conducted to determine risk factors for late mortality, defined as death occurring after hospital discharge and greater than 30 days after surgery. Probability of any RV-PA conduit reintervention was analyzed over time using Fine-Gray modelling.

Results: We reviewed 216 patients, with median follow-up of 2.9 years (range:0.1-8.8). Operative mortaility occurred in 15 patients (7%). Of the 201 survivors, there were 14 (7%) late deaths. DiGeorge syndrome (HR:5.4; 95%CI:1.6-17.8) and need for postoperative tracheostomy (HR:5.9; 95%CI:1.8-19.4) were identified as independent risk factors for late mortality. At least one RV-PA conduit catheterization or surgical reintervention was performed in 109 patients (median time to reintervention:23 months, range:0.3-93). Risk factors for reintervention included use of pulmonary or aortic homografts versus Contegra® bovine jugular vein conduits (HR:1.9; 95%CI:1.2,3.1) and smaller conduit size (HR per mm/m^2:1.05; 95%CI:1.03,1.08).

Conclusions: In a multicenter dataset, DiGeorge syndrome and need for tracheostomy postoperatively were found to be independent risk factors for late mortality after repair of truncus arteriosus, while risk of conduit reintervention was independently influenced by both initial conduit type and size.

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Truncus arteriosus is an uncommon, complex cardiac anomaly that continues to be associated with significant morbidity and mortality beyond the initial surgical repair. In single-center studies with follow-up periods ranging from 2-24 years, mortality beyond the initial surgical hospitalization has been reported to be 2%-15%, while reintervention on the right ventricle to pulmonary artery (RV-PA) conduit later in life is nearly universal and the major source of long-term morbidity.[1-12] While these single-center studies have provided valuable contributions to the literature, they are confounded by center specific surgical and perioperative management strategies, small sample sizes, and data that span different surgical eras

To our knowledge, no multicenter data examining intermediate or long-term outcomes following surgical repair of truncus arteriosus have been published. These data could facilitate risk factor delineation for poor outcome and target high-risk subgroups that deserve increased surveillance beyond the operative period. We therefore aimed use a multicenter dataset to describe early childhood outcomes and determine risk factors for RV-PA conduit reintervention in children who have undergone truncus arteriosus repair in the contemporary era of neonatal repair.

Patients and Methods

Study Population

We retrospectively collected data for children who underwent repair of truncus arteriosus from 2009-2016 at 15 tertiary care referral centers within the United States. A list of participating institutions is provided in Supplemental Table 1. The study was approved by the institutional review boards at all participating centers and was performed in accordance with the ethical standards laid down in the 1964 Declaration of Helsinki and its later amendments. Due to the retrospective nature of the data collected, the need for informed consent was waived.

All patients with truncus arteriosus who underwent initial cardiac repair during the study period were considered for inclusion. Patients with the following criteria were excluded from

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analysis: 1) those who underwent pulmonary artery banding but died prior to repair; 2) diagnosis of hemitruncus or pseudotruncus; or 3) infants who underwent concomitant repair of truncus arteriosus with interrupted aortic arch or aortic arch obstruction (Van Praagh Type A4). Separate analysis of patients with the latter combination of lesions will be reported by this collaboration elsewhere.

Data Collection and Definitions

Baseline characteristics, operative data, and postoperative variables associated with the initial surgical repair were collected for all patients. For operative mortality, we used the current STS-CHSD definition.[13] Data were collected on the occurrence of mortality or RV-PA conduit reinterventions following hospital discharge after the initial surgical repair. Late mortality was defined as mortality after hospital discharge and beyond 30 days of the initial surgical repair. Reintervention was defined as any cardiac catheterization or surgical procedure directed at the RV-PA conduit after the initial surgical repair, which included interventions performed prior to hospital discharge. Data on late mortality and reinterventions after the initial surgical operation were collected on all patients from the time of their surgery to their last documented follow-up appointment.

Statistical Analysis

Data are represented using descriptive statistics, medians with 25th and 75th percentiles for continuous variables and absolute counts with percentages for categorical variables unless otherwise noted. Characteristics of patients who suffered late mortality were compared to patients who survived the initial surgery using Wilcoxon rank-sum, Chi-squared, or Fisher's Exact tests as appropriate for individual variables. A multivariable Cox regression analysis was conducted to determine risk factors for late mortality. Patients who suffered operative mortality were not included in these analyses.

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RV-PA conduits were stratified into categories by indexed conduit diameter and compared using Kaplan-Meier survival analysis. For patients with pulmonary, aortic, and Contegra® RV-PA conduit types, conduit characteristics, follow-up durations, and performance and timing of conduit reinterventions were compared using Chi-squared, Fisher's Exact, or Kruskal-Wallis tests as appropriate. The probability of any RV-PA conduit reintervention was performed using Fine-Gray modelling, which analyzed this outcome over time in the presence of the competing risk of mortality.[14] All statistical analyses were performed using STATA version 14 and SAS version 9.4.

Results

We reviewed 216 patients. Median follow-up was 2.9 years (range: 0.1−8.8). Baseline characteristics and descriptive data from the initial surgical hospitalization are provided in Supplemental Table 2. Genetic testing for 22q.11 deletion was performed in 207 (96%) patients, and 61 (29%) patients were diagnosed with DiGeorge syndrome. Median age at surgery was 10 days (25%,75%: 7,23). Thirty-seven patients (17%) underwent concomitant truncal valve surgery. Postoperative extracorporeal membrane oxygenation (ECMO) was utilized in 10% of the cohort. Operative mortality was 7% (n=15).

Kaplan-Meier survival analysis for all patients is depicted in Figure 1. Of the 201 patients who survived their initial surgical repair, there were 14 late deaths (7%) occurring at a median of 8.3 months (range: 2.5−77.9) after the initial surgery. Ten of the 14 (71%) late deaths occurred within the first 12 months after surgery. Bivariate comparison of patients who died after hospital discharge and those who survived is provided in Tables 1 and 2. DiGeorge syndrome, preoperative mechanical ventilation, performance of an unplanned reoperation excluding postoperative bleeding, and tracheostomy were significantly more common in patients who suffered late mortality, while center volume, concomitant truncal valve surgery at the initial repair and residual truncal valve insufficiency were not associated with late mortality. Using Cox

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Regression analysis, DiGeorge syndrome and tracheostomy at discharge were identified as independent risk factors for late mortality (Table 3). Further details on the events leading to late mortalities are provided in Supplemental Table 3.

One hundred nine patients (50%) underwent at least one RV-PA conduit catheter or surgical reintervention. Median time to first conduit reintervention was 23 months (range: 0.3−93) after initial surgery. Initial conduit intervention was balloon angioplasty in 19 cases, balloon angioplasty with stent placement in 30 cases, placement of a vascular plug for a conduit pseudoaneurysm in one patient, conduit patch in one patient, and replacement in 58 cases. The relationship between conduit size and need for conduit reintervention is illustrated in Figure 2. Patients with smaller conduit diameters placed at their initial surgical repair were more likely to require reintervention during the study follow-up period as compared to patients with larger conduit diameters.

Pulmonary homografts, aortic homografts or Contegra® bovine jugular vein conduits were used for the overwhelming majority (n=191, 88%) of the study population, of which 102 (53%) underwent conduit reintervention. Additional data relevant to patients with one of these conduit types, including the frequency of conduit reinterventions and surgical conduit replacements, are provided in Table 4. Duration of follow-up for patients with each of the three conduit types was statistically similar, but fewer reinterventions were noted in Contegra® bovine jugular vein conduits. Additionally, three cases of endocarditis occurred in our cohort, all of which were due to staphylococcus infection. Two cases occurred in patients that received an aortic homograft and one occurred in a patient with a Contegra® conduit.

Probability of any RV-PA conduit reintervention (e.g., catheter or surgical) or surgical replacement are illustrated in Figure 3 and Supplemental Figure 1, respectively. Probability of conduit reintervention was not statistically different between pulmonary and aortic conduits. Probability of conduit reintervention in pulmonary or aortic conduits at any time point postoperatively however, was significantly greater than that of Contegra® conduits, independent

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of conduit diameter. Independent risk factors for RV-PA conduit intervention included the use of pulmonary or aortic homografts and smaller conduit diameter (indexed to body surface area). Specifically, for every 1 mm/m^2 decrease in RV-PA conduit size, the risk of reintervention increased by 5% ($p<0.001$; Table 5). Similar to our findings regarding late mortality, center volume was not significantly associated with need for RV-PA conduit intervention.

Comment

To our knowledge, this study represents the first multicenter analysis of early childhood outcomes following repair of truncus arteriosus. In our cohort, 14 of the 201 hospital survivors died in the follow-up period. Importantly, the majority (71%) of these deaths occurred within the first year after surgery, a finding that is mirrored by several single-center studies examining outcomes after surgical repair of truncus arteriosus.[1,2,4,10,12,15]

Risk factors for late mortality reported in other studies include severe truncal valve insufficiency, truncal valve replacement, and low weight (≤ 2.5 kg) at the time of initial repair.[3,4,12] These single-center studies included patients who underwent surgery over multiple decades; thus, an "era effect" may have influenced the findings relative to those who underwent surgery during the contemporary era. We identified two independent risk factors for late mortality: DiGeorge syndrome and need for postoperative tracheostomy. Three of 61 patients with DiGeorge syndrome had operative mortality while an additional 10 patients suffered late death, resulting in an overall and late mortality rate of 21% and 17%, respectively. In a single-center retrospective study of 171 patients with truncus arteriosus conducted by Naimo and colleagues, DiGeorge syndrome was also found to be an independent risk factor for late mortality.[12] Unfortunately, further understanding regarding the association between DiGeorge syndrome and late mortality is hindered by the absence of clear explanations for many of the late deaths in our study, which frequently occurred unexpectedly at home or outside hospitals.

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The need for tracheostomy after congenital heart surgery has been associated with poor long-term outcomes in several studies, especially in patients with complex lesions.[16,17] In our analysis, two of twelve patients with tracheostomies died prior to hospital discharge and five suffered late mortality. Data regarding the indications for tracheostomy and details regarding airway anatomy and bronchoscopic findings were not collected, thus making it impossible to determine whether residual cardiac lesions, non-cardiac comorbidities, or a complication related to the tracheostomy itself was the primary cause of death.

Late mortality rates of 17% and 50% for patients with DiGeorge syndrome and tracheostomy, respectively, rival and exceed those observed in patients with hypoplastic left heart syndrome during the interstage period, prior to the widespread adoption of home monitoring.[18] The attention applied to improving interstage mortality in this fragile patient population has led to notable successes via improved surveillance and coordination of care after hospital discharge.[19] Multidisciplinary DiGeorge syndrome follow-up clinics and provision of coordinated post-discharge care could ensure optimal management of non-cardiac comorbidities. Centers should also ensure that their pediatric home ventilation program is in accord with current guidelines.[20] We hypothesize that increased surveillance of high-risk patients with truncus arteriosus could potentially prevent some late deaths with modest additional resource utilization, given the relatively small number of such patients treated at individual centers.

We identified RV-PA conduit size was an independent risk factor for both conduit reintervention and surgical replacement, which is in accord with previous work.[3-5,21,22] Specifically, conduit diameters less than 45 mm/m^2 had the greatest risk of early conduit reintervention. The use of larger conduits aimed at affording patients some degree of freedom from reintervention seems intuitive, but this benefit needs to be reconciled with the possibility that larger conduit size may predispose to greater short-term risk of morbidity and mortality. In a prior analysis of our cohort, we found conduit diameter greater than 50 mm/m^2 to be an

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independent risk factor for major adverse cardiac events (e.g. cardiopulmonary resuscitation, ECMO, or mortality) in the operative period.[23] Based on these data, 45-50 mm/m^2 may represent the conduit diameter range that optimally balances the risk of morbidity and mortality in the immediate postoperative period and long-term complications. Further study should be pursued to confirm these findings. Most importantly, though later and possibly fewer interventions and reoperations are unquestionably desirable, survival after initial repair is paramount and supersedes any efforts to extend the longevity of conduit. Moreover, operative mortality after subsequent RV-PA conduit replacement has been reported to be 0% in numerous contemporary single-center studies.[7,8,21,22]

We also determined that Contegra® bovine jugular vein conduits had significantly lower probability of reintervention than aortic or pulmonary homografts, independent of conduit diameter. This difference was primarily related to differences in the number of cardiac catheterization interventions undertaken in patients with Contegra® conduits as compared to patients with aortic or pulmonary homografts. These findings contrast with several published single-center reports that have attempted to compare the durability of different conduit types, most commonly presented as Kaplan-Meier “conduit survival” analyses or actuarial freedom from reintervention analyses.[4,5,7,8,15,21,22]. These analytical approaches assume that patients who are censored would continue to be at risk of failure events after censorship, which is not the case when patients die prior to conduit intervention. We sought to eliminate this limitation by determining the probability of RV-PA conduit intervention while accounting for the competing risk of mortality. This more rigorous analysis and the multicenter nature of the data, two characteristics which we deem to be strengths of this study, may account for the differences in our findings compared to prior published studies.

This study is limited by its retrospective design. For example, although surviving patients were censored at latest follow-up, data on late mortality or conduit reintervention and replacement for patients who moved or had care transferred to a different institution may not

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have been available and, as a result, occurrence rates for these events may be underestimated. Additionally, echocardiographic or anthropometric data beyond the initial surgical admission was not collected, limiting our ability to assess the potential impact of cardiac performance or somatic growth on outcomes and reinterventions. Lastly, the indication for RV-PA conduit reinterventions and reoperations were not clear in the medical record for most patients and, importantly, likely vary across centers. Accordingly, though we did not find a relationship between center volume and outcomes, it is difficult to disregard a possible overall effect of center on RV-PA conduit reintervention and late mortality from our analyses.

Conclusions

This study represents the first multicenter analysis of early childhood outcomes following repair of truncus arteriosus. Late mortality remains significant in the contemporary era and predominates during the first year after surgical repair. Increased surveillance after hospital discharge for high-risk subpopulations should therefore be considered. Further, use of Contegra® bovine jugular vein RV-PA conduits or conduits with larger diameters may be less likely to require early reintervention, though more study is warranted to determine optimal size and composition of RV-PA conduits to extend conduit longevity without increasing the risk of early morbidity and mortality.

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Table 1. Comparison of Patients with and without Late Mortality after Repair of Truncus Arteriosus – Demographic information and Baseline Patient Characteristics

Variable [a]	Survived (n=187)	Late Mortality (n=14)	*p*-value
Truncus Type (Van Praagh)			0.28
A1	98 (52%)	10 (71%)	
A2	76 (41%)	3 (21%)	
A3	13 (7%)	1 (7%)	
Prematurity (<37 weeks)	33 (18%)	5 (36%)	0.15
Female sex	94 (50%)	7 (50%)	0.98
Chromosomal anomaly, any	69 (37%)	10 (71%)	0.01
DiGeorge/22q.11 deletion	48 (26%)	10 (71%)	0.001
Non-cardiac anatomic anomaly	53 (28%)	6 (43%)	0.36
Prenatal diagnosis	115 (62%)	10 (71%)	0.46
Age at diagnosis (days)	0 (0,2)	0 (0,1)	0.59
Diagnosed before discharge from nursery	147 (79%)	11 (79%)	1.00
Preoperative shock	17 (9%)	2 (14%)	0.63
Preoperative mechanical ventilation	32 (17%)	8 (57%)	0.002
Preoperative infection	24 (13%)	2 (14%)	0.70
Year of surgical repair			0.70
2009 - 2012	90 (48%)	6 (43%)	
2013 - 2016	97 (52%)	8 (57%)	
Center Volume			0.09
< 1 operation / year	15 (8%)	1 (7%)	
≥ 1 < 2 operations / year	20 (11%)	4 (29%)	
≥ 2 < 3 operations / year	103 (55%)	4 (29%)	
≥ 3 operations / year	49 (26%)	5 (36%)	

[a] Continuous variables represented as median (25^{th}%, 75^{th}%); categorical data represented as absolute counts (%)

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Table 2. Comparison of Patients with and without Late Mortality after Repair of Truncus Arteriosus – Data from Initial Surgical Repair

Variable [a]	Survived (n=187)	Late Mortality (n=14)	*p*-value
Age at surgery (days)	10 (7, 25)	7 (6, 14)	0.12
Weight at surgery (kg)	3.1 (2.7, 3.6)	2.8 (2.5, 3.8)	0.46
CPB duration (min)	146 (123, 182)	143 (137, 209)	0.47
Hypothermic circulatory arrest	27 (14%)	4 (29%)	0.24
RV-PA conduit type			0.58
Aortic allograft	47 (25%)	3 (21%)	
Pulmonary allograft	72 (39%)	6 (43%)	
Contegra® conduit	47 (25%)	2 (14%)	
Other / None	21 (11%)	3 (21%)	
RV-PA conduit size (mm/m^2)	51 (45, 55)	51 (46, 59)	0.51
Truncal valve anatomy			0.58
Bicuspid	23 (12%)	3 (21%)	
Tricuspid	100 (54%)	7 (50%)	
Quadricuspid	62 (34%)	4 (29%)	
Dysplastic	2 (1%)	0 (0%)	
Coronary artery abnormality	25 (13%)	0 (0%)	0.14
Truncal valve repaired	27 (14%)	4 (29%)	0.24
Truncal valve replaced	3 (2%)	1 (7%)	0.25
Residual truncal valve insufficiency	95 (51%)	8 (57%)	0.81
Mild	62	5	
Mild-to-moderate	15	2	
Moderate	17	1	
Moderate-to-severe	1	0	
Delayed sternal closure	101 (54%)	11 (79%)	0.07
Postoperative ECMO	11 (6%)	3 (21%)	0.06
Inhaled nitric oxide	86 (46%)	6 (43%)	0.82
Unplanned cardiac catherization	17 (9%)	3 (21%)	0.15
Unplanned reoperation - bleeding	10 (5%)	2 (14%)	0.20
Unplanned reoperation - not bleeding	15 (8%)	4 (29%)	0.03
Tracheostomy prior to discharge	5 (3%)	5 (36%)	<0.001

CPB: cardiopulmonary bypass; RV-PA: right ventricle-to-pulmonary artery; ECMO: extracorporeal membrane oxygenation. Continuous data presented as median (25^{th}%,75^{th}%); categorical data presented as absolute counts (%)

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Table 3. Cox Multivariable Regression Analysis: Risk Factors for Late Mortality

Variable	Hazard Ratio (95% Confidence Interval)	*p*-value
DiGeorge syndrome	5.4 (1.6, 17.8)	0.006
Tracheostomy	5.9 (1.8, 19.4)	0.003
Postoperative ECMO	3.5 (0.9, 13.8)	0.07

ECMO, extracorporeal membrane oxygenation.

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Table 4. RV-PA Conduit Characteristics and Outcomes

Variable [a]	Pulmonary Homograft (n = 83)	Aortic Homograft (n = 53)	Contegra® (n= 55)	*p*-value
Conduit size, mm	10 (9, 11)	9 (9, 11)	12 (12, 12)	<0.001
Conduit size, mm/m^2	50 (44, 54)	49 (46, 55)	54 (48, 58)	0.01
Duration of follow-up, months	36 (18, 60)	36 (22, 63)	42 (15, 77)	0.51
Patients with reinterventions [b]	48 (58%)	33 (62%)	21 (38%)	0.02
Time to first reintervention [b], months	12 (6, 23)	13 (5, 27)	20 (10, 43)	0.28
Patients with surgical replacements	39 (47%)	25 (47%)	19 (35%)	0.29
Time to first replacement, months	23 (8, 37)	26 (11, 31)	20 (11, 55)	0.72

RV-PA, right ventricle-to-pulmonary artery.
[a] Continuous variables represented as median (25th%, 75th%); categorical data represented as absolute counts (%)
[b] Reintervention defined as any cardiac catheterization or surgical procedure directed at the RV-PA conduit after the initial surgical repair, including interventions performed prior to hospital discharge

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Table 5. Multivariable Logistic Regression Analysis of Factors Impacting RV-PA Conduit Durability

Variable	Hazard Ratio	95% Confidence Interval	*p*-value
Probability of Any RV-PA Conduit Intervention			
Pulmonary/Aortic vs. Contegra®	1.9	1.2, 3.1	< 0.01
Indexed Conduit Size (mm/m^2)	1.05	1.02, 1.08	< 0.01
Probability of RV-PA Conduit® Surgical Replacement			
Pulmonary/Aortic vs. Contegra®	1.6	0.96, 2.7	0.07
Indexed Conduit Size (mm/m^2)	1.05	1.02, 1.08	< 0.01

RV-PA: right ventricle to pulmonary artery

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Figure Legends

Figure 1. Kaplan-Meier Survival Curve after Repair of Truncus Arteriosus. Number at risk are provided at each twelve month time point, with number of mortalities during each time interval provided in parentheses. There were 29 deaths, 14 of which were late deaths. The majority (n=10, 71%) of the late mortality occurred within the first 12 months after surgical repair.

Figure 2. Kaplan-Meier curve demonstrating the relationship between conduit size and need for conduit intervention. Need for intervention over time appears to increase as conduit diameter decreases (log-rank p-value < 0.001). Note: Nine patients who did not have conduits placed (i.e. 8 direct anastomoses, 1 systemic-to-pulmonary artery shunt) at the initial surgery are not included.

Figure 3. Probability of RV-PA Conduit Intervention. Contegra® bovine jugular vein conduits (green) had a lower probability of intervention compared with pulmonary (blue) and aortic (red) homografts, accounting for the competing risk of mortality over time (p=0.02).

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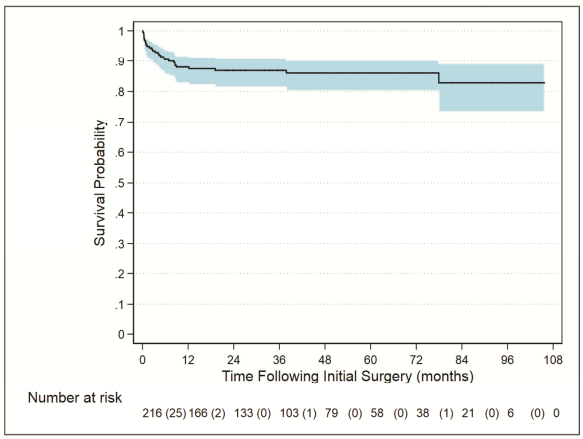

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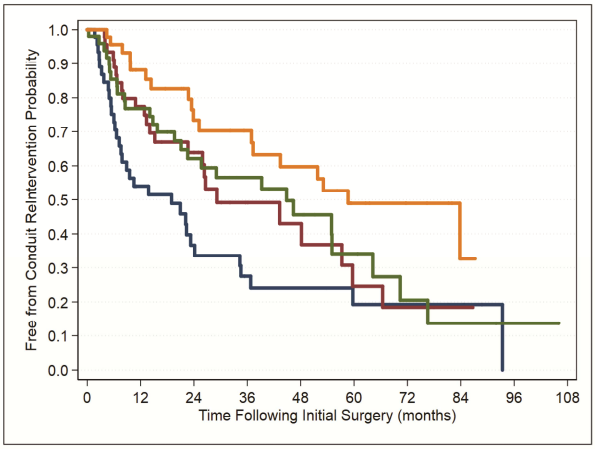

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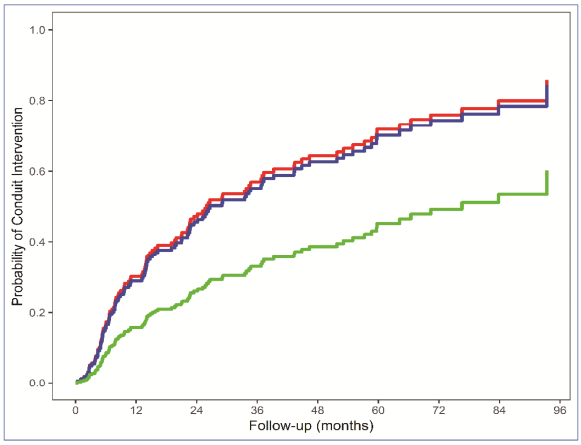

ACCEPTED MANUSCRIPT