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Outcomes of Repair of Common Arterial Trunk with Truncal Valve Surgery: A Review of the STS Congenital Heart Surgery Database

Hyde M. Russell, MD¹, Sara K. Pasquali, MD², Jeffrey P. Jacobs, MD³, Marshall L. Jacobs, MD⁴, Sean M. O'Brien, PhD⁵, Constantine Mavroudis, MD⁴, and Carl L. Backer, MD¹

¹Division of Cardiovascular-Thoracic Surgery, Children's Memorial Hospital, Department of Surgery, Northwestern University Feinberg School of Medicine, Chicago, IL

²Department of Pediatrics, Duke University School of Medicine and Duke Clinical Research Institute, Durham, NC

³The Congenital Heart Institute of Florida, Division of Thoracic and Cardiovascular Surgery, All Children's Hospital and Children's Hospital of Tampa, University of South Florida College of Medicine, St. Petersburg, FL

⁴Department of Pediatric and Congenital Heart Surgery, Cleveland Clinic, Cleveland, OH

⁵Department of Biostatistics, Duke University School of Medicine, Durham, NC

Abstract

Background—Previous studies have suggested truncal valve insufficiency may adversely affect outcome after common arterial trunk (CAT) repair. It is unknown if truncal insufficiency requiring truncal valve surgery (TVS) at the time of primary CAT repair impacts outcome.

Methods—Patients in the STS Congenital Heart Surgery Database undergoing CAT repair from 2000–2009 were included. Outcomes were compared for those with and without TVS at the time of CAT repair and were further stratified by interrupted aortic arch (IAA) repair.

Results—Of 572 patients (63 centers), median age at surgery was 12d (interquartile range 6–34d). Twenty-three patients underwent concomitant TVS (n=22 repair, n=1 replacement) during CAT repair, and 4 patients underwent TVS later during the same hospitalization (n=1 repair, n=3 replacement). Thirty-nine patients underwent IAA repair at the time of CAT repair, 5 of whom had concomitant TVS. Mortality for CAT repair with TVS vs. isolated CAT repair was 30% vs. 10% (p=0.0002). All 4 patients who required TVS later during the admission died. TVS was associated with increased mortality in CAT patients both with and without IAA repair, with the highest mortality (60%) in CAT patients undergoing IAA repair and TVS (n = 5). CAT + TVS had an increased risk of mechanical support and a longer hospital stay.

Conclusions—TVS in patients undergoing CAT repair is associated with significant mortality. IAA repair and TVS at the time of CAT repair carries particularly high risk. Failure to address significant truncal insufficiency, necessitating early reoperation with TVS, had uniformly poor outcomes.

Correspondence to: Hyde M. Russell, MD, Division of Cardiovascular-Thoracic Surgery, Children's Memorial Hospital, 2300 Children's Plaza, mc 22, Chicago, IL 60614, hrussell@childrensmemorial.org. Office: 773-880-4378, Fax: 773-880-3054.

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Keywords

congenital heart disease; truncus; truncal valve; database; outcomes; pediatrics; surgery; incisions; exposure; techniques

Introduction

Common arterial trunk (previously referred to as *truncus arteriosus*) is a rare congenital heart defect defined by the presence of a solitary great artery that exits the heart through a common ventriculo-arterial junction and supplies directly the systemic, pulmonary, and coronary arterial pathways [1–3]. Repair of this defect involves separating the systemic and pulmonary pathways with establishment of a connection between the right ventricle and the pulmonary arteries and closure of the ventricular septal defect. Patients with common arterial trunk were historically treated with delayed complete repair, sometimes preceded by palliation with banding of the pulmonary arteries [4]. For the past two decades or more, most patients have been managed by early primary repair, predominately as neonates [5].

The truncal valve in patients with common arterial trunk may have two, three, or four leaflets and exhibit varying degrees of regurgitation. It is unclear whether truncal valve regurgitation of a severity mandating truncal valve repair or replacement impacts outcomes of patients undergoing common arterial trunk repair. Multiple studies have identified preoperative truncal valve regurgitation as a risk factor for mortality [6–9]. Others, however, have not found the presence of truncal valve insufficiency nor the requirement for truncal valve intervention to impact outcomes [10–12]. We undertook this study to evaluate the prevalence and impact of truncal valve surgery on postoperative outcomes in patients undergoing common arterial trunk repair in a large multi-center cohort.

Material and Methods

The Society of Thoracic Surgeons (STS) Congenital Heart Surgery Database was utilized for this study. This database contains preoperative, operative, and postoperative outcomes data on all children undergoing heart surgery at participating centers. It is the largest existing clinical congenital heart surgery data registry in the world, representing nearly three quarters of all U.S. centers performing congenital heart surgery [13]. Data quality and reliability are assured through intrinsic verification of data and a formal process of site visits and data audits [14]. The Duke Clinical Research Institute serves as the data warehouse and analysis center for all of the STS National Databases. Research performed on the STS database at the Duke Clinical Research Institute is reviewed by the Duke University Institutional Review Board. Because the data used for research represent a limited data set (no direct patient identifiers) that was originally collected for non-research purposes, and the investigators do not know the identity of individual patients, the analysis of these data was declared by the Duke IRB to be research not involving human subjects. This study was also reviewed and approved by the STS Access and Publications Committee.

Patient Population

Patients undergoing primary common arterial trunk repair at any of the 85 U.S. centers submitting data to the STS Congenital Heart Surgery Database from 2000–2009 were eligible for inclusion. A total of 77 centers performed at least 1 repair of common arterial trunk during this time period. While the STS Database contains nearly complete data for the required standard data fields regarding procedure and in-hospital mortality, not all centers submit complete data for the other variables in the STS Database. Therefore it is standard practice to exclude centers with >15% missing data for key study variables, in order to

maximize data integrity and minimize missing data. This left 63 centers in the final study population.

Data Collection

Data collection included patient characteristics of age, weight, weight-for-age z-score, gender, and any non-cardiac abnormality as defined in the STS Database [15]. Operative characteristics included cardiopulmonary bypass time and aortic cross clamp time. Data regarding truncal valve surgery (repair or replacement) either at the time of common arterial trunk repair or later during the same admission were collected, along with whether the patient underwent concurrent interrupted aortic arch repair at the time of common arterial trunk repair.

Outcomes—The primary outcome was in-hospital mortality. Secondary outcomes included utilization of postoperative mechanical circulatory support, and postoperative length of stay.

Statistical Analysis—Study variables were described using standard summary statistics. The prevalence of truncal valve surgery at the time of initial common arterial trunk repair or later during the same admission was described. Patient characteristics and outcomes were compared in those undergoing common arterial trunk repair and truncal valve surgery vs. isolated common arterial trunk repair using Wilcoxon rank sum and chi-square tests adjusting for within-center clustering, and stratified by interrupted aortic arch repair. Within the truncal valve surgery group, patient characteristics were compared in those who survived to hospital discharge vs. those who died inhospital using the Wilcoxon rank sum and chi-square tests. Due to the small number of patients and events, multivariable analyses were not possible. All analyses were performed using SAS version 9.2 (SAS Institute Inc, Cary, NC). A p-value <0.05 was considered statistically significant.

Results

Patient population

A total of 572 patients undergoing common arterial trunk repair were included (Figure 1). Twenty-seven patients (5%) underwent 29 truncal valve related procedures during primary common arterial trunk repair or later during the same hospitalization. Twenty-three patients underwent truncal valve surgery at the time of initial common arterial trunk repair (22 valve repairs and 1 valve replacement). Two patients underwent truncal valve surgery later during the same admission after isolated common arterial trunk repair (both valve replacements, performed 7 and 61 days after initial common arterial trunk repair). Two patients underwent truncal valve repair at the time of initial common arterial trunk repair and then had subsequent truncal valve surgery later during the same admission (one with redo truncal valve repair 85 days after initial surgery, and one with truncal valve replacement 36 days after initial surgery). Thirty-nine patients underwent interrupted aortic arch repair at the time of common arterial trunk repair, 5 of whom required truncal valve surgery.

Patient characteristics

Table 1 displays patient and operative characteristics for the overall cohort, and in those undergoing common arterial trunk repair + truncal valve surgery vs. those undergoing isolated common arterial trunk repair. Overall, median age at surgery was 12 days (interquartile range 6d to 34d), median weight was 3.2 kg (interquartile range 2.7–3.6 kg), 50% were male, and 43% had a non-cardiac abnormality. Patients in the common arterial trunk repair + truncal valve surgery group underwent repair earlier compared to patients undergoing isolated common arterial trunk repair (7 d vs 13 d, $p = 0.03$). There were non-

significant trends toward fewer non-cardiac abnormalities and a greater proportion with concurrent interrupted aortic arch repair in the common arterial trunk + truncal valve surgery group. Operative data were notable for longer cardiopulmonary bypass times and aortic cross clamp times in the valve group, as expected.

Center characteristics

Patients from 63 centers participating in the STS database were included in this study. Centers with greater than 15% key variables missing (n=14) were excluded from the study. Median annualized volume among the 63 centers was 403 cases per year. The distribution of valve repairs performed by center volume quartile is presented in Table 2. This analysis did not reveal any bias toward or away from truncal valve repair based on center volume. The small number of patients undergoing truncal valve surgery precluded meaningful analysis of the relationship of center volume with outcome (n = 23).

Outcomes

Overall, 61 of the 572 patients in the cohort (11%) died in-hospital (Table 3). Of note, all four patients who underwent later valve surgery during the same hospital admission died (Figure 1). Truncal valve surgery was associated with increased mortality in patients with common arterial trunk both with (n=5) and without (n=22) interrupted aortic arch repair, with the highest mortality in patients with common arterial trunk undergoing interrupted aortic arch repair and truncal valve surgery (60%). There were no other differences in preoperative or operative variables between survivors and non-survivors.

Postoperative mechanical circulatory support was utilized in 45 patients (7.8%) in the overall cohort, and was significantly more common in the truncal valve surgery group vs. the isolated common arterial trunk repair group (18% vs. 7%, p=0.002). Postoperative length of stay was also longer in the common arterial trunk + truncal valve surgery group (Table 3).

Comment

This study utilized the STS Congenital Heart Surgery Database to assess outcomes of patients with common arterial trunk and truncal valve insufficiency, who underwent truncal valve repair or replacement during the same hospital admission as their primary common arterial trunk repair. We found that patients undergoing truncal valve surgery had a significantly higher mortality when compared to patients undergoing isolated common arterial trunk repair. This risk was magnified in the presence of interrupted aortic arch. Patients requiring subsequent reoperation for truncal valve repair or replacement during the same hospital admission had a uniformly poor outcome.

Risk factors for poor outcome have been analyzed since the first reported repair of common arterial trunk by McGoon, Rastelli, and Ongley [4]. The association of truncal valve insufficiency with mortality risk has been noted by multiple authors. Ebert's report in 1984 of 100 patients with common arterial trunk undergoing repair in the first six months of life revealed a mortality rate of 11%, remarkably similar to the overall mortality rate in this study. In that series 8 of the 11 deaths had evidence of preoperative truncal valve insufficiency; one of whom underwent truncal valve replacement [5]. Hanley and colleagues published a series of 63 patients undergoing repair of common arterial trunk at Children's Hospital Boston. They found severe truncal valve insufficiency prior to surgery was a significant risk factor for early death in both univariate and multivariate analyses. In addition, Hanley reported on the long term follow-up of patients undergoing repair of common arterial trunk in infancy and found severe truncal insufficiency to be a risk factor for late death among initial hospital survivors (n = 27, 30% mortality [6]. Di Donato and

colleagues from the Mayo Clinic found moderate or severe truncal valve regurgitation to be associated with poor long term survival in a study of 167 patients over a 17 year period (n = 62, 50% mortality) [8]. Pearl and associates at UCLA reviewed their experience of 32 patients who underwent common arterial trunk repair in infancy and similarly concluded that truncal valve insufficiency is an incremental risk factor for early and late mortality [9].

In contrast to these reports, some recent single center series have not shown truncal valve regurgitation to be associated with increased mortality at the time of primary repair. Bove and associates reported a series of 46 neonates undergoing repair of common arterial trunk at the University of Michigan. Five patients required truncal valve replacement but neither truncal valve regurgitation nor truncal valve replacement was related to death [10]. A recent report by Hawkins and associates showed that in their institution's experience (n = 42), complex cases with either truncal valve insufficiency, interrupted aortic arch or both were associated with greater utilization of resources but not with a higher rate of operative mortality [16]. Valve repair techniques have evolved with the hope of mitigating the negative effect of truncal insufficiency on outcomes. There are multiple single center series of patients with common arterial trunk who underwent concomitant repair of the common arterial trunk and truncal valve surgery with good short term results [17–22].

The observation that certain centers have “neutralized” the risk associated with severe truncal valve insufficiency may or may not be generalizable. There has been no large scale multi-institutional report of outcomes following repair of common arterial trunk prior to this study. The Congenital Heart Surgeons' Society published a report of 50 neonates with combined interrupted aortic arch and common arterial trunk. In that cohort the overall mortality was 68% and severe truncal valve regurgitation was found to be uniformly fatal [23]. The present study corroborates these results as we found a similar rate of death (60%) among patients who underwent both repair of interrupted aortic arch and truncal valve surgery in conjunction with repair of common arterial trunk. It has been speculated that the physiologic derangement from residual aortic obstruction following interrupted aortic arch repair is magnified in the setting of a regurgitant truncal valve [24].

Our study identified three populations of patients undergoing common arterial trunk repair who had a significantly increased risk of operative mortality. Patients undergoing truncal valve surgery had a three-fold greater risk of mortality compared with those who did not require valve intervention. The higher utilization of mechanical circulatory support and longer length of stay attests to the increased complexity of these patients. Concomitant interrupted aortic arch repair also had a significantly higher risk of mortality in patients undergoing common arterial trunk repair. The combination of interrupted aortic arch and truncal valve regurgitation requiring truncal valve surgery, while rare (occurring in 5 patients), was associated with particularly high mortality (60% vs. 9% if neither condition existed). Despite making up only 6% of the overall cohort, patients in these three subgroups accounted for a quarter of the overall mortality.

The small cohort of patients requiring reoperation during the same hospitalization for truncal valve repair (n=1) or replacement (n=3) is of particular interest, as these patients had uniformly poor outcomes. Two patients in this reoperative cohort required postoperative mechanical assistance after their first operation indicating a compromised hemodynamic state. These four patients returned to the operating room between 7 and 85 days postoperatively for valve surgery. Although multiple factors may have contributed to the fatal outcomes of these patients, there is nonetheless concern that failure to address significant truncal insufficiency during the first operation may be an important predictor of death.

Limitations

This study is limited by the unaccounted variables that may have affected the results of this retrospective review. In particular, degree of truncal valve regurgitation, valve morphology and technique of repair, and postoperative valve function are not known. Furthermore, the patients were analyzed not by the presence of truncal valve insufficiency, but by the occurrence of truncal valve surgery. The true denominator of patients with significant truncal valve regurgitation could not be ascertained from the database, therefore truncal valve surgery is used as a surrogate for this condition. Selection bias for truncal valve intervention by the operating surgeon could also certainly affect these results. Although this study represents the largest analysis of outcomes in patients undergoing common arterial trunk repair, it was also limited by the small number of patients and events which precluded more in-depth analysis beyond descriptive evaluation.

Conclusions

Centers caring for neonates and infants with common arterial trunk have achieved good outcomes despite its rarity. This multi-center study suggests that the overall mortality rate currently is comparable to the best results published 20 years ago, despite nearly all operations now being done in the neonatal period and the inclusion of more complex patients who may not have survived to operation in previous eras. The subgroups of increased complexity identified in this study will demand heightened attention and investigation in order to continue to improve outcomes in this patient population in the future.

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Figure 1.

Patient Population

CAT, common arterial trunk, IAA, interrupted aortic arch, TVS, truncal valve surgery

Table 1

Preoperative and Operative Data

Variable	Overall (n = 572)	Isolated CAT Repair (n = 545)	CAT Repair + TVS (n = 27)	p-value
<i>Patient Characteristics</i>				
Age (days)	12 (6, 34)	13 (6, 35)	7 (5, 11)	0.03
Male gender	288 (50%)	277 (51%)	11 (41%)	0.45
Weight (kg)	3.2 (2.7, 3.6)	3.2 (2.7, 3.6)	3 (2.7, 3.4)	0.62
Non-cardiac abnormality	244 (43%)	237 (43%)	7 (26%)	0.20
<i>Operative Data</i>				
Concurrent IAA repair	39 (7%)	34 (6%)	5 (19%)	0.07
CPB time (min)	149 (117, 183)	148 (117, 180)	192 (145, 232)	0.02
Cross clamp time (min)	83 (66, 87)	82 (65, 106)	106 (81, 140)	0.008

CAT, common arterial trunk; CPB, cardiopulmonary bypass; IAA, interrupted aortic arch; TVS, truncal valve surgery.

Data are displayed as frequency (percent) and median (interquartile range).

Table 2

Distribution of Valve Repairs

Mean Annualized Center Volume	Valve Repair Frequency	Percent
1 st Quartile	5	22%
2 nd Quartile	8	35%
3 rd Quartile	3	13%
4 th Quartile	7	30%
Total	23	100%

Median annual volume of all included centers was 404 cases per year.

1st quartile represents the lowest volume centers and 4th quartile represents the highest volume centers.

Table 3**Outcomes**

	Overall (n = 572)	Isolated CAT Repair (n = 545)	CAT Repair + TVS (n = 27)	p-value
<i>In-Hospital Mortality</i>				
Overall	61 (11%)	53 (10%)	8 (30%)	0.0002
Without IAA repair	50 (9%)	45 (9%)	5 (23%)	0.003
With IAA Repair	11 (28%)	8 (24%)	3 (60%)	0.41
Post-op Mechanical Circulatory Support	45 (8%)	40 (7%)	5 (18%)	0.002
Post-op Length of Stay (days)	16 (10, 29)	15.5 (10, 29)	29 (14, 83)	0.02

CAT, common arterial trunk; IAA, interrupted aortic arch; TVS, truncal valve surgery

Data are displayed as frequency (percent) and median (interquartile range).