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Truncus Arteriosus Associated with Interrupted Aortic Arch in 50 Neonates: A Congenital Heart Surgeons Society Study

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Background. Patients with both interrupted aortic arch (IAA) and truncus arteriosus (TA) have worse outcomes than those with either lesion in isolation. We determined outcomes and associated factors in this rare group.

Methods. From 1987 to 1997, 50 (11%) of 472 neonates with IAA were identified with TA. Site of aortic arch interruption was distal to the left subclavian artery in 16% and between the left common carotid and subclavian artery in 84%. From the common arterial trunk, the pulmonary arteries arose from a main pulmonary trunk in 46%, common orifice in 22%, and separate orifices in 32%. At presentation, truncal valve stenosis was present in 12% and regurgitation in 22%.

Results. There were 34 deaths, with a single early hazard phase. Overall survival from admission was 44%, 39%, and 31% at 6 months, 1 year, and 10 years, respectively. One patient had primary cardiac transplantation

and 4 died without any intervention. The IAA repair alone was performed in 7 patients, with single stage repair of both IAA and TA in 38 patients. Associated factors for overall time-related death include female gender ($p < 0.001$), type III TA ($p < 0.001$) and one institution (low-risk; $p < 0.001$). Results improved somewhat over time ($p < 0.001$). At 5 years after IAA repair only 28% were alive without arch repair intervention, and at 5 years after TA repair only 18% were alive without conduit reoperation.

Conclusions. The combination of IAA and TA carries high early mortality, with high risk of reinterventions in survivors. One stage repair of both TA and IAA is the optimal management.

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The truncus arteriosus communis (TA) is an uncommon cardiac anomaly with an incidence of approximately 0.7% of congenital heart disease [1]. Aortic arch interruption (IAA) is found in approximately 15% of these children [2, 3], making the combination of these anomalies rare. Gomes and McGoon [4] performed the first successful repair of the combined lesions in a 2-year-old patient in 1971. Although a few successful repairs have been reported during the last three decades, the cumulative experience with this rare combination in neonates is limited to case reports and two small series of 7 and 9 patients, respectively [5–14]. We therefore sought to determine the characteristics, management, and outcomes of this group of patients in a prospective multi-institutional study of 50 neonates.

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Patients and Methods

From 1987 to 1997, 472 neonates with IAA were prospectively enrolled from 33 institutions of the Congenital Heart Surgeons Society (CHSS), 50 (11%) of whom had associated TA. The number of patients enrolled per institution ranged from 1 to 8, with five institutions enrolling 4 or more. All consented patients with IAA admitted to a CHSS institution within 30 days of birth were eligible for inclusion. Participation by member institution was voluntary and confidential, and ethical approval was obtained as per local requirements. Ethics approval for the CHSS Data Center is obtained annually from the Research Ethics Board of the Hospital for Sick Children, Toronto.

Definitions

CLASSIFICATION. To accurately describe truncus morphology of our patients we used a modification of Collett and Edwards original classification [15] (Fig 1). The Van

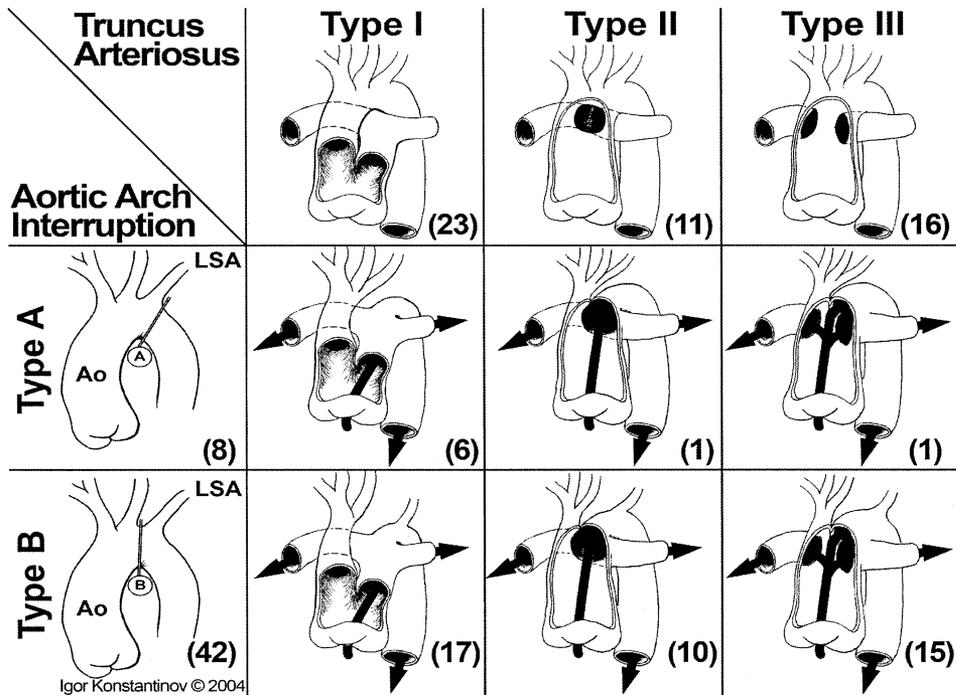


Fig 1. Anatomy of the aortic arch interruption and truncus arteriosus in 50 neonates. (Ao = aorta; LSA = left subclavian artery.)

Praagh classification was not used because all of our patients had one or another variation of type A4 [1–3]. Type I TA is defined as a clearly defined main pulmonary trunk giving rise to both left and right pulmonary arteries; type II as TA with minor separation of the pulmonary artery orifices; type III as TA with pulmonary arteries originating from opposite sides of a common arterial trunk, or as one of the pulmonary arteries supplied from the ductus (thus including type A3 of Van Praagh's classification); and type IV as TA with both pulmonary arteries originating from the descending aorta. None of our patients were classified as type IV TA.

Aortic arch interruption was defined as either a complete discontinuity or a nonpatent fibrous strand in the transverse arch or aortic isthmus. IAA types were described using Celoria and Patten classification [16] (Fig 1). Patients with right aortic arch and other associated aortic arch anomalies were individually described.

TRUNCAL VALVE PATHOLOGY. Truncal valve insufficiency was classified as mild, moderate, or severe as determined upon initial admission. Truncal valve stenosis was classified as mild (mean transvalvular gradient ≤ 20 mm Hg), moderate (mean gradient 20–50 mm Hg), and severe (mean gradient ≥ 50 mm Hg). Truncal valve morphology was described as number of cusps of the common truncal valve.

OUTFLOW OBSTRUCTION. Obstruction across the arch anastomosis was defined as a mean pressure gradient of 30 mm Hg or greater by transthoracic echocardiography. Obstruction of the right ventricle to pulmonary trunk was defined as a gradient of two-thirds or more of systemic pressure.

Data Collection

Data were abstracted from submitted copies of medical records requested for initial and subsequent assessments, admissions, and procedures. Given variability in the standardization and comprehensiveness of submitted echocardiographic reports, we adapted an echocardiographic measurement protocol for TA associated with IAA from our previously described protocol for patients with left heart obstructive lesions [17]. Initial (before any important intervention) echocardiogram videotape recordings were requested from contributing institutions. The submitted recordings were then reviewed and standardized measurements made by a single experienced pediatric echocardiographer (ASM) blinded to subsequent management and outcome of the patient. From an initial study population of 50 patients, 21 initial echocardiogram recordings were received and reviewed. Data were abstracted from detailed echocardiography reports regarding an additional 23 patients for whom a recording was not submitted for review. Echocardiographic-derived cardiac structural variables, including left ventricular dimensions, were converted into Z-scores using regression equations based on previously published nomograms [18, 19]. The most recent cross-sectional follow-up was performed between March and June 2004. Follow-up is available for 12 of 16 presumed current survivors.

Data Analysis

Goals of the analysis were to (1) describe morphologic, procedural, and institutional characteristics, and (2) determine incremental risk factors for time-related mortality, reinterventions directed at arch repair ob-

Table 1. Patient Characteristics (n = 50)

Variable	Value	Missing	Deaths (n = 34)
<i>Demographic characteristics</i>			
Age at admission (days, median, range)	4 (birth–30)	0	
Gender (female/male)	28:22	0	23:11
Birth weight (kg, median, range)	2.9 (1.9–4.9)	14	
Weight at first operation (kg, median, range)	3.0 (1.8–4.7)	12	
< 2.0 kg at first surgery	1 (3%)	12	1
2.1–3.0 kg at first surgery	19 (50%)	12	12
> 3.0 kg at first surgery	18 (47%)	12	8
Age at first operation (days, median, range)	9 (1–48)	1	
Prematurity (< 37 weeks EGA)	7 (18%)	10	7
Noncardiac anomaly	7 (14%)	0	6
DiGeorge anomaly	7 (39%)	32	7
<i>Predominant presenting symptoms</i>			
Congestive heart failure	14 (34%)	9	9
Cyanosis	11 (27%)	9	4
Murmur	4 (10%)	9	2
Tachypnea	4 (10%)	9	4
Acidosis	3 (7%)	9	2
Necrotizing enterocolitis	2 (5%)	9	2
Irritability	1 (2%)	9	1
Other	3 (7%)	9	2
<i>Morphologic characteristics</i>			
<i>Type IAA</i>			
Type A	8 (16%)	0	4
Type B	42 (84%)	0	30
<i>Type TA</i>			
Type I	23 (46%)	0	12
Type II	11 (22%)	0	9
Type III	16 (32%)	0	13
<i>TV Insufficiency</i>	24 (53%)	5	18
Mild	13 (29%)		9
Moderate	7 (16%)		5
Severe	4 (9%)		4
<i>TV Stenosis</i>	17 (38%)	5	13
Mild	11 (24%)		8
Moderate	4 (9%)		3
Severe	2 (4%)		2
Peak instantaneous gradient (mm Hg, median, range)	29 (10–93)	32	
Ascending aortic diameter (mm, mean \pm SD)	4.5 \pm 1.2	28	
Truncal annulus diameter (mm, mean \pm SD)	11.2 \pm 1.5	25	
Truncal root diameter (mm, mean \pm SD)	13.8 \pm 2.8	28	
LPA Z-score (median, range)	0.8 (–1.3–4.0)	23	
RPA Z-score (median, range)	0.8 (–1.1–1.8)	23	
MPA diameter (mm, mean \pm SD)	11.9 \pm 1.7	34	
Mitral valve Z-score (median, range)			
Anterior-posterior dimension	1.1 (0.8–1.3)	33	
Lateral dimension	1.1 (0.8–1.3)	32	
Any mitral valve regurgitation	5 (14%)	15	
Any mitral valve stenosis	1 (3%)	19	
<i>Cardiovascular anomalies</i>	15 (30%)	0	12
Hypoplastic left heart syndrome	1 (2%)		0
HRV, azygos continuation of the IVC	1 (2%)		1
Aberrant subclavian artery	7 (14%)		5

Table 1. (Continued)

Variable	Value	Missing	Deaths (n = 34)
Left superior vena cava	2 (4%)		2
Multiple VSDs	2 (4%)		2
Malalignment VSD	8 (16%)		5
Right aortic arch	3 (6%)		2
TAPVD	1 (2%)		1
<i>Preoperative Management Characteristics</i>			
Intubated	23 (62%)	13	12
Use of prostaglandin	29 (100%)	21	13
Use of dopamine	12 (41%)	31	8

EGA = estimated gestational age; HRV = hypoplastic right ventricle; IAA = interrupted aortic arch; IVC = inferior vena cava; LPA = left pulmonary artery; MPA = main pulmonary artery; RPA = right pulmonary artery; TA = truncus arteriosus; TAPVD = total anomalous pulmonary venous drainage; TV = truncal valve; VSD = ventricular septal defect

struction after IAA repair, and conduit reinterventions after TA repair using competing risk methodology. Data are given as frequency, median with range, or mean ± SD as appropriate, with the number of non-missing values indicated. Data analyses were performed with SAS statistical software (version 9; SAS Institute, Inc, Cary, NC). Continuous variables were compared with unpaired two-tailed *t* tests. Multiphase parametric modeling of the hazard function and competing risks methodology were used to define rates of transition to mutually exclusive time-related events and incremental risk-factors associated with each outcome as previously described [20].

Results

Prevalence and Patient Characteristics

Truncus arteriosus with associated IAA was found in 50 (11%) of 472 neonates enrolled neonates. All patients had a large malalignment ventricular septal defect (VSD) and patent ductus arteriosus. Patient and morphologic characteristics of these 50 neonates are shown in Table 1. Predominant presenting symptoms included congestive heart failure in 14 patients, cyanosis in 11 patients, tachypnea or an isolated murmur in 4 patients each, and metabolic acidosis in 3 patients. Two patients had grade III intraventricular hemorrhage and 2 patients presented with necrotizing enterocolitis requiring laparotomy. Preoperative management consisted of prostaglandin infusion in 100%, mechanical ventilation in 62%, and inotropic support in 42%. Gestational age was known in 40 patients, and prematurity (defined as < 37 weeks gestation) was present in 7 neonates, 6 of whom died. Antenatal diagnosis was not documented in any patients. A flow chart of events after admission is shown in Figure 2.

Associated Anomalies

CARDIOVASCULAR ANOMALIES. Seven (12%) neonates had associated cardiac anomalies, including multiple VSDs (n = 2), hypoplastic left heart physiology (n = 1), obstructed supracardiac total anomalous pulmonary venous drainage (n = 1), left superior vena cava (n = 2), and hypo-

plastic right ventricle with azygos continuation of the inferior vena cava (n = 1). The presence of these anomalies was universally fatal, except one with hypoplastic left heart physiology who underwent heart transplantation. AORTIC ARCH ANOMALIES. Right aortic arch was present in 3 (6%) neonates, one of whom had an aberrant subclavian artery. Six patients had left aortic arch with aberrant subclavian artery.

TRUNCAL VALVE. Truncal valve regurgitation was found in 24 (48%) patients, 18 of whom died. All 4 patients with severe regurgitation died. Seven patients had moderate regurgitation, 5 of whom died. There were 9 deaths among the 13 patients with mild regurgitation. Truncal valve stenosis was found in 16 (32%) patients, 13 (81%) of whom died. All patients (n = 2) with severe stenosis died, 3 of 4 patients with moderate stenosis died, and 8 of 10 patients with mild stenosis died. The combination of both truncal valve regurgitation and stenosis was found in 12 patients, with 9 deaths.

Associated Noncardiac Anomalies

Noncardiac anomalies were present in 6 patients: hydrocephalus (n = 1), cleft palate (n = 3), and tracheoesophageal fistula (n = 2), one of whom also had associated mesomelic dysplasia that consisted of bilateral absence of the thumb

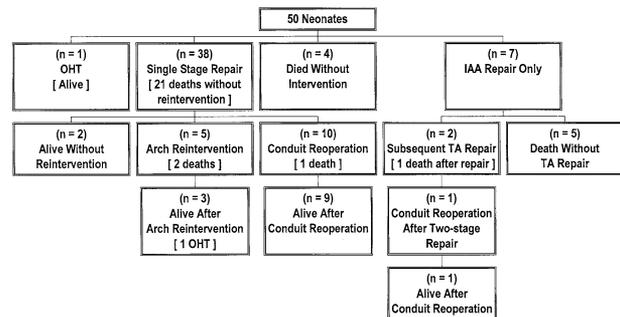


Fig 2. Flow chart illustrating events after admission in 50 neonates with truncus arteriosus (TA) and interrupted aortic arch (IAA). (OHT = orthotopic heart transplantation.)

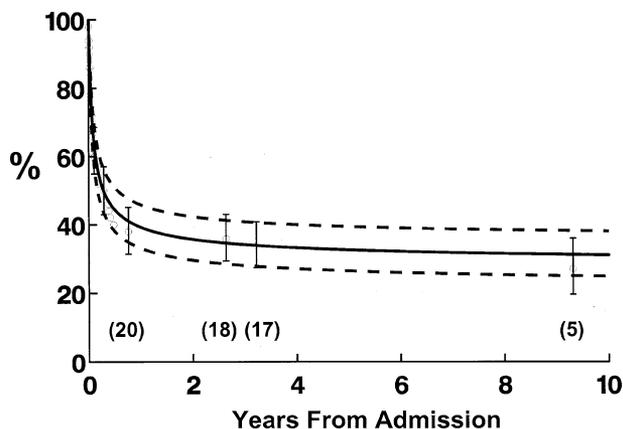


Fig 3. Overall time-related survival of 50 neonates with truncus arteriosus and interrupted aortic arch. All patients began at the time of initial admission to a CHSS member institution. Solid lines represent parametric point estimates; dashed lines enclose 70% confidence interval; circles with error bars represent nonparametric estimates; numbers in parentheses indicate the number of patients at risk. (Overall survival: 6 months, 44%; 1 year, 39%; 10 years, 31%.)

and radius. Two patients underwent tracheoesophageal fistula repair after cardiac repair. All 6 patients died.

DIGEORGE SYNDROME. DiGeorge syndrome was documented in 10 (20%) patients. Type B IAA was present in 9 of them. Truncus arteriosus was type I in 4, type II in 3, and type III in 3. Aberrant subclavian artery was present in 4 patients with DiGeorge syndrome. Six patients in this group died, including all 4 patients with aberrant subclavian artery. Two patients with DiGeorge syndrome developed necrotizing enterocolitis, including one who received palliative care only and one who underwent complete repair with homograft replacement of the truncal root, but died during the same admission.

Surgical Repair

Median age at operation was 9 days (range, 1-48 days) and median weight was 3.0 kg (range, 1.8-4.7 kg).

ONE STAGE REPAIR. The TA and IAA were repaired at the same time in 38 patients, 18 (47%) of whom died during the same admission.

TWO STAGE REPAIR. The IAA was repaired first in 7 patients, including 5 patients who died without TA repair. Truncus arteriosus was repaired as a second stage procedure during the same admission in the remaining 2 patients, one of whom died 4 days after TA repair. Operative mortality for those patients who underwent staged repair was 86%.

OTHER TYPES OF REPAIR. One patient with hypoplastic left heart physiology, who remains alive, underwent Norwood palliation followed by heart transplantation.

TYPE OF IAA REPAIR. The IAA was repaired by direct anastomosis in 91%, 48% of whom had concomitant patch augmentation. Interposition graft was used in 9%.

TRUNCAL VALVE PROCEDURES. Truncal valve procedures were performed in 7 patients, including truncal root replacement with aortic allograft in 3 patients. Two were performed during the initial operation with one death. One truncal valve replacement was performed as a late reoperation, and this patient remains alive. Truncal valve repair was performed during the initial operation in 3 patients, all of whom died. One patient had later truncal valve repair and remains alive. Patients with type III TA had longer cardiopulmonary bypass times (271 ± 57 min) than neonates with other truncal types (139 ± 55 min), $p = 0.007$.

Overall Mortality

There were 34 (68%) deaths with a single early hazard phase (Fig 3). No operations were done in 4 (8%) patients, all of whom died (parents refused surgery for 2 patients, one deemed inoperable, one died of cardiac tamponade during diagnostic catheterization). Causes of death are listed in Table 2. Overall survival from admission was 44% at 6 months, 39% at 1 year, and 31% at 10 years. Incremental risk factors for time-related death include female gender ($p < 0.001$), type III truncus arteriosus ($p < 0.001$), and one institution (low-risk; $p < 0.001$). Results improved over time ($P < 0.001$) (Table 3).

The favorable influence of later birth cohort on overall survival from admission is shown in Figure 4. The multivariable hazard model for mortality was solved for three different birth cohorts (1988, 1993, 1997), using birth cohort as the sole predictor of mortality. While survival improved with successive birth cohorts, the amount of improvement was less for more recent patients.

Aortic Arch Related Reinterventions After Repair

Of the 46 patients who had repair of IAA either in isolation (7 patients) or combined with TA repair (38 patients), this was followed by death in 24 patients and survival to a reintervention for post-IAA repair anastomotic obstruction in 7 patients. Of the 7 arch reinterventions 4 were reoperations, consisting of patch augmentation

Table 2. Causes of Death ($n = 34$)

In-Hospital Deaths	28
Died without operation	4
Intraoperative deaths	3
Death on ECMO	3
Postoperative cardiac failure	12
Sepsis	3
Complications after repair of tracheoesophageal fistula	1
Unknown	2
Late Deaths	6
Refractory seizures (date unknown)	1
Chronic rejection 3 years after OHT	1
Sudden death at 4.5 months	1
Complications of abdominal surgery 9 years after repair	1

ECMO = extracorporeal membrane oxygenation; OHT = orthotopic heart transplantation.

Table 3. Incremental Risk Factors for Time-Related Death (n = 34), With and Without Institutions as Additional Risk Factors

Variable	Parameter Estimate (±SE)	p Value	Reliability (%)
a. Without institutions			
Earlier date of birth ^a	0.67 ± 0.20	<0.001	83%
Female gender	1.19 ± 0.37	0.001	87%
Type III truncus	0.81 ± 0.36	0.03	60%
b. With institutions			
Earlier date of birth ^b	0.63 ± 0.19	<0.001	83%
Female gender	1.50 ± 0.38	0.001	87%
Type III truncus	1.25 ± 0.38	0.001	60%
Not in institution A ^b	1.7 ± 0.63	0.006	64%

^a Entered after inverse transformation. ^b One institution was found to be lower risk (labeled A).

SE = standard error.

of the arch, one was a transcatheter balloon dilatation of an anastomotic stenosis, and 2 had an initial balloon dilation followed by subsequent patch augmentation.

The time-related hazard function for death without an IAA repair intervention was characterized by an early phase. The hazard function for survival to an IAA repair intervention was also characterized by a single early phase, accounting for seven events. The competing risks for the three mutually exclusive events: death without an IAA repair intervention, survival to an IAA repair intervention, and remaining alive without an intervention showed that after 5 years, 58% had died without an IAA repair intervention, 28% remained alive without an IAA repair intervention, and 14% had an IAA repair intervention (Fig 5).

Conduit Related Reinterventions After TA Repair

The IAA and TA were repaired as a single-stage procedure in 38 patients, and repair of TA followed initial IAA repair in 2 additional patients. The TA repair was followed by death in 25 patients, and survival to reintervention on either the right ventricle-pulmonary artery con-

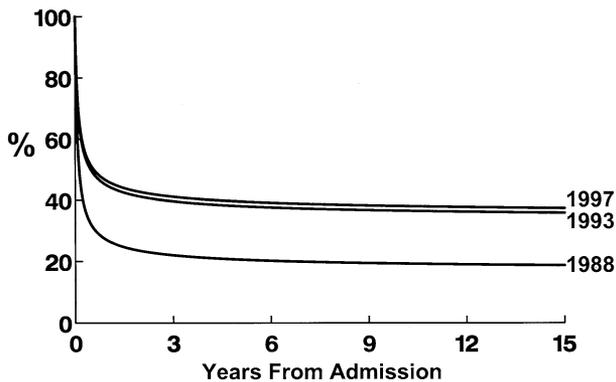


Fig 4. Predicted overall survival for the first 10 years after admission stratified by birth cohort.

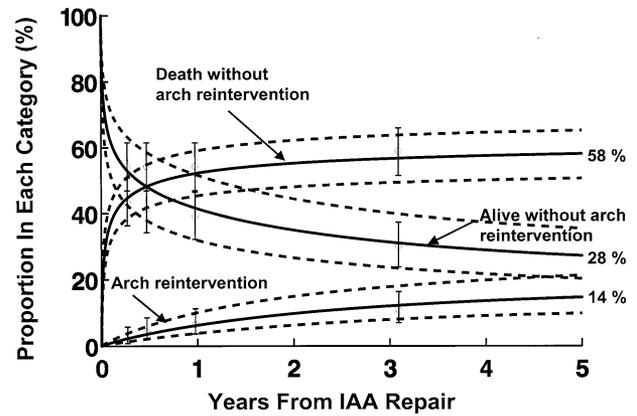


Fig 5. Competing risk depiction for events after initial repair of interrupted aortic arch (IAA). All patients began at the time of initial IAA repair (n = 46), and could transition to either death or a subsequent procedure aimed at addressing potential or actual arch obstruction. Solid lines represent parametric point estimates; dashed lines enclose 70% confidence interval; circles with error bars represent nonparametric estimates. Estimated proportion of patients (expressed as percentage of total) in each of 3 categories at 5 years after repair are shown on right.

duit, or the pulmonary trunk conduit, in 12 patients. Reoperation was performed in 11 patients, conduit replacement for stenosis in 9, and conduit patch enlargement in 2 patients. Six patients who had conduit reoperation also had at least one balloon dilatation of the pulmonary arteries, and 5 had subsequent aortic arch reintervention.

The hazard function for time-related transition to subsequent right ventricle-pulmonary trunk conduit reoperation was characterized by a prolonged late hazard phase, accounting for 11 events. The hazard function for transition to death without a subsequent conduit reoperation was characterized by a steep early hazard phase, consisting of 16 events, and a prolonged late hazard phase, with 9 events. The competing risks for the two events showed that after 5 years after TA repair, 59% had died without conduit reoperation, 23% had conduit reoperation, and 18% were surviving without conduit reoperation (Fig 6).

Comment

In this study, the outcomes and associated risk factors for death, arch repair reintervention, and conduit reoperation are described for the largest current cohort of neonates with TA in the setting of IAA.

Overall Mortality

In our series, a slight improvement over time was noted in overall survival, although it remained poor even for patients born in 1997. In our recent report of 472 neonates with interrupted aortic arch, overall survival was 60% at 10 years after admission, including the 50 patients with associated TA [21]. The competing risk analysis performed in the previous report consistently associated the

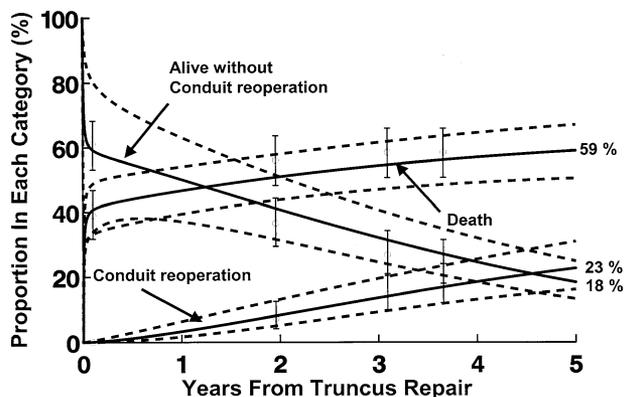


Fig 6. Competing risks depiction of events after truncus repair. All patients began at the time of initial truncus repair ($n = 41$), and could transition to either death or conduit reoperation. Solid lines represent parametric point estimates; dashed lines enclose 70% confidence interval; circles with error bars represent nonparametric estimates. Numbers in parentheses at right indicate the estimated prevalence in each of the states at 5 years.

presence of TA with increased mortality [21]. Other identified risk factors for mortality included IAA type B or C, younger age at study entry, lower birth weight, and female gender [21]. In the present analysis, overall mortality was associated with TA with the pulmonary arteries arising separately (type III), earlier birth cohort, female gender, and one institution (low-risk).

Type III TA may require unifocalization of the pulmonary arteries, which necessitates a more complex and lengthy operative procedure [22]. Patients with this morphology had longer cardiopulmonary bypass times than neonates with more favorable anatomy. In addition, the presence of pulmonary vascular disease, though not directly assessed in this analysis, is increased in this group of patients, particularly if definitive repair was delayed. Currently, the technique of truncal transection with harvest of a generous truncal "cuff" containing the pulmonary artery orifices is the preferred approach, and should lead to more favorable outcomes in patients with discontinuous pulmonary arteries. Type III TA has not been previously reported as an incremental risk factor for mortality, likely owing to the paucity of this subset in previous reports [13, 14, 22]. Two retrospective series, however, have documented the clinical importance of TA in general [22, 23]. Two of the 4 late deaths in a series of 63 patients with TA, by Hanley and colleagues [23], occurred in patients with discontinuous pulmonary arteries.

Female gender was identified as a risk factor for death in both the IAA cohort [21], and the present series. Numerous studies of adult cardiac patients have demonstrated the unfavorable influence of female gender on outcome [24–26]. However, it remains unclear whether gender per se, the existence of referral bias, or the presence of unmeasured covariates actually contribute to the increased risk in females [25, 27]. The emergence of gender as a risk factor cannot be explained by the current report.

Improved outcomes in one institution are related to patient factors. No patients treated in this institution had moderate or severe truncal regurgitation, and none had type III TA. We did not identify any differences in management characteristics in this institution, and single stage versus two-stage approach was employed with equivalent frequency in this institution compared with the 5 other centers enrolling more than 4 patients.

IAA as a Risk Factor

We have previously reported that patients with TA and IAA have worse outcomes than those patients with isolated IAA [21]. The presence of significant left ventricular outflow tract obstruction may increase the degree of truncal valve regurgitation and accelerate the appearance of congestive heart failure. Maintenance of a balanced circulation preoperatively is also more difficult in those with TA and IAA. Myocardial ischemia is potentiated by reduced diastolic coronary flow and subendocardial coronary perfusion is reduced by volume load-induced increases in left-ventricular end-diastolic pressure. In contrast, others have reported the potential neutralization of associated IAA as a risk factor for adverse outcome after TA repair [13, 14]. However, important differences between the study populations exist. Jahangiri and colleagues [14] analyzed the results of surgical repair of TA in 50 children operated from 1992 to 1998. Nine patients, including 7 neonates, had associated IAA, and none had truncal valve regurgitation. In this series, there were no deaths in the 9 patients with TA and associated IAA at 3-year follow-up and none required reoperation directed at the truncal valve or reintervention for important arch obstruction. Similarly, Sano and colleagues [13] reported 7 patients with IAA and TA that underwent complete one stage repair of both anomalies between 1985 and 1989. All patients had either type I or type II TA. Although no data on truncal valve regurgitation or stenosis are available in this report, none of 7 patients required intervention on the truncal valve. There were no early or late deaths at mean follow-up of 29 months (range, 5 months to 4 years) from the initial repair, and reoperations were performed in only 3 patients.

In contradistinction to the above studies, 48% of patients in the current series had truncal valve regurgitation with or without concomitant truncal valve stenosis, and an additional 8% had isolated stenosis. Although truncal valve status (regurgitation or stenosis) did not emerge as a statistically significant risk factor, the prevalence of death tended to be higher for those with greater than mild functional impairment. Further, the true impact of truncal valve regurgitation on overall mortality may be underestimated in the current analysis because the functional status of the valve was obtained preoperatively, when runoff to the pulmonary circulation may conceal important valvar regurgitation.

Truncal Valve Intervention

Early phase mortality predominated in infants after single stage repair. The majority of patient deaths resulted

from myocardial failure, and occurred during initial hospitalization. Current techniques employed during single stage repair include relief of arch obstruction, most commonly by direct anastomosis with or without patch augmentation, and placing a conduit to restore right ventricle-pulmonary trunk continuity. Because results with early truncal valve replacement have been unsatisfactory, truncal valve replacement is avoided except when severe regurgitation mandates intervention [14, 28, 29]. Truncal valve repair has emerged as a more favorable option, obviating the need for repeated replacement in infants undergoing rapid somatic growth [22, 30, 31]). In the present study, the number of patients who underwent repair was inadequate to extrapolate as to which anatomic substrate would be most amenable to valvuloplasty techniques.

Severe truncal valve regurgitation was universally fatal in our population. That the majority of patient deaths were attributable to low cardiac output suggests that improved truncal valve function may mitigate against early cardiovascular compromise, especially when poor preoperative status is present. Several studies [14, 22, 23, 30, 32] have indeed shown a survival disadvantage in patients with residual truncal regurgitation after single stage repair. Patients with important truncal valve regurgitation should therefore be managed aggressively. Early heart transplantation should be considered if reasonable truncal valve function cannot be achieved with valvuloplasty techniques.

Aortic Arch Reinterventions

Recurrent aortic arch obstruction occurred in 7 patients, and tended to be related to initial repair by methods other than direct anastomosis with patch augmentation. This agrees with our findings in the previous IAA report [21], where direct anastomosis, augmented with either allograft or pericardium, provided the most durable repair. Patients who underwent patch augmentation at the time of arch reintervention have not required subsequent procedures for recurrent obstruction, whereas 2 of those who had transcatheter relief of obstruction had subsequent patch repair. It is also possible that initial arch augmentation may reduce truncal valve incompetence by preventing subsequent outflow tract obstruction.

Inferences and Recommendations

Despite continuing improvement in outcomes in the modern era, survival for TA in the presence of IAA is poor. The optimal management of complex TA is evolving, and considerable debate exists with regard to type of initial arch repair and the approach to the functionally impaired truncal valve. Although the heterogeneity of this lesion mandates individualization of management, our data suggest that early single stage repair using direct anastomosis with patch augmentation is the optimal approach. Low cardiac output was responsible for the majority of deaths in this series, and thus strategies to improve postoperative myocardial performance are warranted. Aggressive management with truncal valve re-

pair, when feasible, or early heart transplantation should be considered in those infants where truncal valve repair is not possible.

Summary

The combination of TA and IAA carries a very high early mortality, with an important risk of reinterventions in survivors. Patients with associated noncardiac anomalies and functional truncal valve impairment have poor prognosis. Outcomes may be improved with single stage repair of both lesions. Patch augmentation of the IAA is the optimal management strategy.

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INVITED COMMENTARY

Truncus arteriosus associated with interrupted aortic arch (TA-IAA) is a rare disease. This Congenital Heart Surgeons Society (CHSS) multi-institutional retrospective cohort of 50 patients undergoing surgical intervention for TA-IAA is the largest series ever published [1]. Previous single institution studies have been done on less than 10 patients. The high mortality reported (61% mortality at 1 year) contrasts with the excellent results obtained when the truncus arteriosus is isolated. Furthermore, several centers [2-4] have reported that TA associated with IAA and TA associated with truncal valve regurgitation can be safely managed.

The reasons for the negative results reported by this CHSS study are multifactorial. The surgical outcome of TA-IAA is highly dependant on the preoperative status of the patient. The truncal valve regurgitation can be particularly severe in TA-IAA. If there is no data to suspect that the truncal valve is more malformed when associated with an IAA, any truncal regurgitation will be worsened by the premature closure of the ductus arteriosus, which induces an increase of the pulmonary flow and therefore of the truncal valve flow. Any delay in the institution of PGE1 may induce myocardial ischemia, NEC, and ultimately multiple organ failure. These complications are prevented by early PGE1 infusion, minimal FiO₂, normal to elevated PCO₂ (similar to HLHS management) and also very early repair in first days of life and fetal diagnosis. This multicentric series reports a median age at surgery of 9 days, which is rather late.

The risk of the TA-IAA repair is increased by noncardiac factors. The presence of either a prematurity or DiGeorge syndrome or an extracardiac anomaly is asso-

ciated in this study with 100% mortality. All the severe risk factors are currently defined in the comprehensive Aristotle score as shown by a recent study on TA-IAA from Miyamoto and colleagues [5].

The questionable surgical technique applied to many of the patients in this series is a cause of failure as acknowledged by the authors. The optimal technique of TA-IAA repair was simultaneously published in the mid 1990s by Hanley and colleagues [4], Jahangiri and colleagues [3], Bove and colleagues [6], and us [7]. This technique includes one-stage repair in the first days of life, mobilization of the descending aorta, direct aortic anastomosis, transection of the common trunk, harvesting of a large pulmonary artery branches cuff, patch enlargement of the ascending aorta, and valved conduit reconstruction of the RVOT. An ascending aorta patch enlargement is essential [7] to prevent ascending aorta stenosis due to the mismatch between the truncal root and the small diameter of the ascending aorta, particularly in the presence of type B with aberrant right subclavian artery.

The authors are following the 1949 classification from Collet and Edwards [8]. In our opinion, the 1987 modified classification [9] from Van Praagh that excluded the Collet-Edwards type III is far more realistic. The pulmonary artery (PA) branches arise usually in a one and a half motion and do not raise a real technical challenge because we divide the truncus root and harvest a large PA branch cuff. The real difficulty is coming from the PA branches arising from the descending aorta or from a ductus-like vessel (type A3 of Van Praagh, not mentioned in this series).

This study proposed a heart transplant for the most critical forms associated with severe aortic regurgitation.