Canadian Journal of Cardiology 35 (2019) 446–452

Clinical Research

Contemporary Outcomes and Factors Associated With Mortality After a Fetal or Postnatal Diagnosis of Common Arterial Trunk

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See editorial by Hancock Friesen and Jaquiss, pages 376–378 of this issue.

ABSTRACT

Background: Common arterial trunk (CAT) is a rare anomaly with a spectrum of pathology. We sought to identify current trends and factors associated with postnatal outcomes.

Methods: This was a single-centre review including 153 live births with planned surgery. Patients were analyzed as 2 cohorts based on era of CAT diagnosis (1990 to 1999 vs 2000 to 2014) and complexity of disease (simple vs complex). "Complex" required the association with significant aortic arch obstruction, truncal valve (TV) stenosis/regurgitation, and/or branch pulmonary artery (PA) hypoplasia, respectively.

Results: Sixteen (10%) died preoperatively, and this outcome was associated with significant TV stenosis (odds ratio [OR] 4.55; $P = 0.01$) and regurgitation (OR 3.17; $P = 0.04$); 130 (95%) of 137 operated infants underwent primary complete repair. Their survival rates to 1 year improved from 54% to 85% after 2000, although this outcome remained substantially lower for cases with a complex vs simple CAT repair (76% vs 95%; OR 6.46; $P = 0.006$). Other risk factors associated with decreased 1-year survival included diagnosis before 2000 (OR 4.48; $P = 0.038$) and a lower birth weight (OR 8.0 per kg weight; $P = 0.001$). Finally, of 93 survivors beyond year 1 of life, 76 (82%) had undergone a total of 224 reinterventions. Only 15 (16%) were alive without any surgical or catheter-based reintervention at study end.

RÉSUMÉ

Contexte : Le tronc artériel commun (TAC) est une anomalie rare associée à un large éventail de maladies. Dans notre étude, nous avons tenté de recenser les tendances actuelles et les facteurs ayant un lien avec l'issue postnatale.

Méthodologie : Cette étude monocentrique a porté sur 153 bébés nés vivants pour lesquels une intervention chirurgicale était prévue. Les patients ont été répartis en deux cohortes en fonction de la période pendant laquelle le diagnostic de tronc artériel commun avait été posé (de 1990 à 1999 vs de 2000 à 2014) et de la complexité de l'anomalie (simple vs complexe). Elle était considérée comme « complexe » si elle était associée à une obstruction importante de la crosse de l'aorte, à une sténose ou à une insuffisance de la valve troncale, ou à une hypoplasie d'une branche du tronc pulmonaire.

Résultats : Seize (10 %) nouveau-nés sont morts avant d'avoir été opérés. Ces décès ont été associés à une sténose (risque relatif approché [RRA] : 4,55; $p = 0,01$) et à une insuffisance (RRA : 3,17; $p = 0,04$) importantes de la valve troncale; 130 (95 %) des 137 nouveau-nés opérés ont subi une réparation initiale complète. Après l'an 2000, le taux de survie à 1 an a grimpé de 54 à 85 %, bien que ce pourcentage soit resté considérablement plus faible dans les cas de réparation d'un TAC complexe que dans les cas simples (76 % vs 95 %;

Common arterial trunk (CAT) is a rare form of congenital heart disease (CHD), in which a single arterial trunk gives rise to the aorta and the pulmonary arteries. Complete repair during the neonatal period is the management of choice.[1] In most patients, this includes the use of some sort of right ventricle (RV) to pulmonary artery (PA) conduit. Others may also require surgical reconstruction of an obstructed or interrupted aortic arch (IAA), repair or replacement of the truncal valve (TV), and arterioplasty of hypoplastic and/or disconnected branch pulmonary arteries. Although operative results continue to improve, CAT remains associated with significant morbidity and mortality. This includes the need of reinterventions for conduit obstruction, branch PA stenosis, and TV pathology.[2-6] Moreover, outcome assessments usually do not include patients who were not offered surgery or have died while awaiting surgery. On the other hand, CAT is increasingly detected before birth.[7,8] This study sought to examine our single-centre experience with CAT since 1990 and to identify markers predictive

Received for publication October 22, 2018. Accepted December 4, 2018.

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See page 451 for disclosure information.

https://doi.org/10.1016/j.cjca.2018.12.006

Conclusions: Despite recent surgical improvements, postnatal mortality continues to be substantial if CAT is complicated by significant pathology of the TV, aortic arch, or branch PAs. Reoperations and catheter interventions are eventualities for most patients during childhood.

RRA : 6,46; $p = 0{,}006$). Les autres facteurs de risque liés à une baisse du taux de survie à 1 an comprenaient les suivants : diagnostic posé avant l'an 2000 (RRA : 4,48; $p = 0{,}038$) et un plus petit poids à la naissance (RRA : 8,0 par kilogramme; $p = 0{,}001$). Enfin, sur les 93 bébés ayant survécu à leur 1re année de vie, 76 (82 %) avaient subi, au total, 224 nouvelles interventions. À la fin de l'étude, seulement 15 enfants (16 %) étaient toujours en vie sans avoir eu besoin d'une nouvelle intervention chirurgicale ou d'un cathétérisme.

Conclusions : Malgré les avancées réalisées récemment en chirurgie, la mortalité postnatale reste élevée dans les cas où le TAC se complique d'affections majeures de la valve troncale, de la crosse de l'aorte ou d'une branche du tronc pulmonaire. La plupart des patients risquent de devoir subir de nouvelles chirurgies et des interventions par cathétérisme pendant leur enfance.

of mortality. We postulated that improved prenatal detection and perinatal care of CAT have yielded an overall reduction in postnatal mortality in the current era.

Methods

The institutional Research Ethics Board approved this retrospective study (REB #1000017862).

Patients

The Hospital for Sick Children provides exclusive tertiary care for children with complex CHD in Ontario (~140,000 live births per year). Included were referrals from within our province with a diagnosis of CAT from January 1990, to December 2014. A modified classification by Collett and Edwards[9] was used to categorize each case into one of 3 possible subtypes. In type I, the main PA originates from the anterior/lateral aspect of the common trunk and then branches. Type II is defined by separate but adjacent origins of the branch PAs from the posterior/lateral aspect of the trunk. In type III, the branch PAs originate apart from the lateral aspects of the trunk. The rare subtype IV, now considered a form of pulmonary atresia, was excluded.[10] Patients who did not have their primary operations at The Hospital for Sick Children or patients followed at different institutions were excluded.

Data collection

Collected patient data included demographics, spectrum of abnormalities, management, and outcome to December 31, 2016. Echocardiographic findings were retrieved offline from recordings and reports. The diagnosis of mild, moderate, and severe TV stenosis was based on Doppler flow peak velocities between 2.6 and 2.9 m/s, 3.0 and 4.0 m/s, and > 4 m/s, respectively, in the absence of ventricular dysfunction. TV regurgitation was graded as absent, mild ($< 25\%$), moderate (25% to 64%), or severe ($\geq 65\%$) from the width of the central color Doppler jet.[11]

Definitions and outcome measures

Demographic, echocardiographic, and operative parameters were analyzed for correlation with mortality and surgical or catheter-based reinterventions. Operative death was defined as death within 30 days from the time of surgery. Patients were analyzed as 2 cohorts: live births diagnosed from 1990 to 1999 (era 1) vs 2000 to 2014 (era 2) and live births undergoing a primary complete repair of a simple vs complex form of CAT. The distinction between eras reflects the significant changes in prenatal CAT diagnosis and operative techniques during these periods (see *Discussion* section). We mainly used RV-PA homografts in era 1 and Contegra bovine grafts (Medtronic, Minneapolis, MN) in era 2. Complex CAT was defined by the presence of at least 1 of these lesions: aortic arch obstruction, moderate or severe TV regurgitation and/or stenosis, and hypoplastic and/or discontinuous branch PAs.[12]

Statistical analysis

Data were summarized by frequencies, median with range or mean ± standard deviation, as appropriate. Between-cohort differences in continuous and dichotomous/polytomous variables were evaluated using Wilcoxon rank sum and Fisher's exact tests, respectively. Variables with low-frequency categories were reported in the descriptive statistics but collapsed (when possible) or excluded for further analyses. Kaplan–Meier estimates were used to quantify mortality and log-rank test to assess between-cohort differences. For cases with a primary biventricular repair, cumulative proportion of patients with reinterventions since the primary repair was calculated, using death without repeat interventions as a competing risk. Associations between death with risk factors were assessed and quantified in terms of odds ratios (ORs) and 95% confidence intervals (CIs) using logistic regression. Multivariable models included clinically relevant factors. Mean imputation was used to account for missing variables. Statistical analyses were performed with R v3.4.1 and SAS v9.4 (SAS Software, Cary, NC).

Results

Inclusion criteria were fulfilled by 165 cases, including 49 fetuses and 116 infants referred at 22 (16 to 35) weeks of gestation and 5 (1 to 310) days of life, respectively. The proportion of fetal CAT diagnosis was 6% (4 of 64 cases) in era 1 (1990 to 1999) and 45% (45 of 101) in era 2 (2000 to 2014; $P < 0.001$). There was a steady increase in prenatal detection rates of CAT during era 2 (2000 to 2004: 28%; 2005 to 2009: 50%; 2010 to 2014: 63%; $P = 0.03$).

Table 1. Characteristics of all fetal and infant cases at the time of CAT diagnosis (n = 165) and at birth (n = 153) summarized by era

	All cases		Live births				Era 1 vs 2
			Era 1		Era 2		*P* values
Number of cases	165		64	42%	89	48%	
Time of diagnosis:							< 0.001
Fetal	49		4	6%	33	37%	
Postnatal	116		60	94%	56	63%	
Truncus type:							0.10
Type 1	101	61%	34	53%	61	69%	
Type 2	56	34%	28	44%	24	27%	
Type 3	8	5%	2	3%	4	4%	
Truncus valve regurgitation:							0.03
None to mild	129	78%	44	69%	75	84%	
Moderate to severe	36	22%	20	31%	14	16%	
Truncus valve stenosis:							1.00
None to mild	137	83%	53	82%	73	82%	
Moderate to severe	28	17%	11	17%	16	18%	
Aortic arch obstruction:	30	18%	11	17%	19	21%	0.54
Interrupted	27		10		17		
Coarctation	3		1		2		
Pulmonary arteries:							
Hypoplastic and/or discontinuous	25	15%	13	20%	9	10%	0.10

Spectrum of congenital abnormalities

Table 1 summarizes the characteristics of all patients and all live births by era. Relevant other cardiac anomalies included multiple ventricular septal defects (n = 9), right aortic arch (n = 44), total anomalous pulmonary venous connections (n = 3), AV valvar atresia (n = 2), and major coronary anomalies (n = 2; anomalous left coronary artery from the pulmonary artery [ALCAPA]; intramural right coronary artery [RCA]). Genetic disorders were documented in 51 of 129 (40%) cases undergoing testing, including microdeletion 22q11 (n = 39), trisomy 21 (n = 2), trisomy 13 (n = 1), various duplication anomalies (n = 7), del 8p23 (n = 1), and balanced translocation of chromosomes 2 and 17 (n = 1). Other associations included VACTERL (n = 4); Goldenhar syndrome (n = 1); McKusick-Kaufman syndrome (n = 1); and various craniofacial (n = 8), abdominal (n = 6), limb/vertebral (n = 6), and genital (n = 2) anomalies.

Preoperative presentation, management, and outcomes

Termination of pregnancy was elected for 11/49 (22%) fetuses; the fetus with trisomy 13 died spontaneously. The median age and weight at birth of the 153 live-born cases was 38 (25 to 42) gestational weeks and 2.86 (0.46 to 4.7) kg, respectively. Compared with the postnatally diagnosed group, the live-born fetal cohort did not differ in the severity of cardiac pathology, postnatal management, and outcome (data not shown) but was born significantly earlier (37 vs 39 gestational weeks, $P < 0.001$), with a lower birth weight (2.56 vs 2.87 kg, $P = 0.01$).

Sixteen (10%) live-born infants died prior to surgery at a median age of 8 (1 to 47) days. Reasons of preoperative death included multiorgan failure (n = 9), cardiac arrest (n = 1), bowel ischemia with perforation (n = 3), respiratory failure (n = 2), and extreme prematurity complicated by sepsis (n = 1). Preoperative death by univariate logistic regression was significantly associated with a fetal diagnosis (9 of 37 vs 7 of 116 postnatal; OR 5.01; CI, 1.71-14.61; $P < 0.001$), moderate/severe TV regurgitation (7 of 16 vs 27 of 137 survivors to surgery; OR 3.17; CI, 1.08-9.27; $P = 0.04$), moderate to severe TV stenosis (7/16 vs 20/137; OR 4.55; CI, 1.52-13.61; $P = 0.01$), the need of preoperative ventilation (14 of 16 vs 52 of 137; OR 11.44; CI, 2.50-52.40; $P < 0.001$) and inotropic support (13 of 16 vs 19 of 137; OR 26.91; CI, 7.01-103.39; $P < 0.001$), but not with study era (era 1: 7 of 64 vs era 2: 9 of 89; OR 1.09; CI, 0.38-3.1; $P = 1.00$).

Surgical management and results

Surgery (n = 137) consisted either of a primary complete repair (n = 130), staged complete repair (n = 3), heart transplant (n = 2), or univentricular surgery (n = 2).

Primary repair. Median age at surgery was 13 days with no difference between eras (era 1 vs 2: 10 (2 to 336) vs 14.5 (2 to 88) days; $P = 0.63$). Nonetheless, infants with simple CAT underwent surgery significantly later than those with complex CAT (21 [2 to 338] vs 7 [2 to 271] days; $P < 0.001$). The RV outflow was reconstructed in 76 (58%) cases with a homograft, in 44 (34%) with a Contegra bovine graft, in 6 (5%) with a polystan conduit, and in 4 (3%) with a direct RV-PA anastomosis. The median conduit diameter was 12 (7 to 16) mm. The aortic arch was concurrently repaired in 26 (20%) and the TV repaired or replaced in 17 (13%). Median cardiopulmonary bypass time was 127 minutes (era 1 vs 2: 153 (63 to 445) vs 115 (67 to 365) minutes; $P = 0.0013$) and median aortic cross-clamping time 74 minutes (era 1 vs 2: 59 (12 to 160) vs 83 (14 to 179) minutes; $P = 0.0001$).

Figure 1A shows freedom from death estimates by era. Median patient follow-up was 8.6 (0.01 to 19.7) years. Survival rates at 1 month, 1 year, and 10 years before 2000 were 66%, 54%, and 50% and improved to 93%, 85%, and 81%, respectively, since 2000. Similarly, the operative mortality decreased from 36% to 7% since 2000 (era 1 vs 2: 20 of 56 vs 5 of 74; $P < 0.0001$). Figure 1B compares the survival of simple vs complex CAT since 2000: 10-year survival was 95% with a simple CAT when compared with 68% if CAT were complex. Factors associated with decreased 1-year survival are

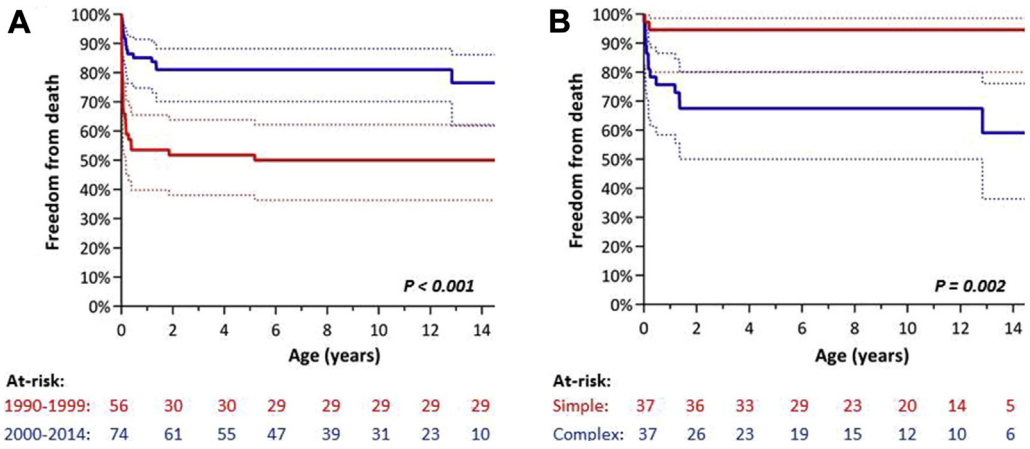

Figure 1. Freedom from death estimates of patients (**A**) with a primary complete repair of CAT by era and (**B**) with a primary repair of a simple (**red**) vs complex (**blue**) CAT between 2000 and 2014 during era 2. **Dotted lines** represent 95% confidence intervals of the mean.

listed in Table 2. The same variables were also significantly associated with operative death (not shown).

Nonprimary repair. Four (57%) of 7 children without primary biventricular repair were alive at study end (age range: 5.4 to 16 years), including 1 of 2 with single ventricle palliation, 1 of 2 with a heart transplant, and 2 of 3 with staged biventricular repairs.

Reoperations

The cumulative proportions of patients requiring TV surgery and repeat interventions of any type, catheter-based procedures, and conduit surgery following a primary CAT repair are shown in by era Figure 2A-D. Of 93 survivors beyond year 1 of life, 76 (82%) had undergone a total of 224 (1 to 8) reinterventions by the time of the last visit (n = 71; age: 13.8 [1.2 to 19.7] years) or death (n = 5; age: 5.2 [1.2 to 16.5]) years. Procedures included new or repeat TV surgery in 18 patients (19%; 11 repairs, 7 replacement; patient age: 7.3 [1.16 to 16.1] years), RV-PA conduit replacement in 53 (57%; 68 surgeries; time to first reoperation: 4.6 [0.09 to 13.4] years) as well as catheter interventions for conduit/PA stenosis in 68 (73%; 131 procedures; time to first procedure: 0.9 [0.01 to 13.2] years) and pulmonary valve implants in 7 (8%; 11.2 [7.9 to 16] years). At study end, only 15 patients (16%; age 2.3 to 13.5 years) were alive without any reintervention.

Table 2. Factors associated by univariable and multivariable regression analysis with decreased survival to 1 year of life in patients undergoing primary biventricular repair of common arterial trunk (CAT; N = 130)

	N	Death	N	Alive	Univariable OR [95% CI]	*P* value	Multivariable OR [95% CI]	*P* value
Years of diagnosis	37		93					
• < 2000 (Era 1)		26		30				
• ≥ 2000 (Era 2)		11		63	0.20 [0.085-0.45]	< 0.001	0.22 [0.05-0.89]	0.038
CAT type	37		93					
• Type 1		23 (62%)		59 (63%)	1.06 [0.47- 2.31]	0.60	0.96 [0.29-3.19]	0.95
• Type 2 or 3		14 (38%)		34 (37%)				
Fetal diagnosis	37	5 (14%)	93	22 (24%)	0.50 [0.16-1.36]	0.20	0.36 [0.06-1.89]	0.25
Prematurity ≤ 35 weeks	36	7 (19%)	85	9 (11%)	2.18 [0.72- 6.37]	0.155	0.53 [0.05-0.89]	0.53
Weight at birth (kg)	36	2.75 (1.0-4.3)	85	2.88 (1.5-4.4)	0.54 [0.28- 0.99]	0.051	0.13 [0.03- 0.38]	0.001
Male gender	37	14 (38%)	93	40 (43%)	0.81 [0.36-1.75]	0.59	1.09 [0.36-3.29]	0.88
Chromosomal anomalies	26	12 (46%)	77	31 (40%)	1.27 [0.52-3.12]	0.60	1.02 [0.19-4.99]	0.98
Complex CAT	37	11 (30%)	93	54 (58%)	3.27 [1.48-7.65]	0.004	6.46 [1.82- 27.30]	0.006
• AAI or coarctation	37	10 (27%)	93	16 (17%)	1.78 [0.71-4.37]	0.21		
• Moderate/severe TV regurgitation	37	10 (27%)	93	16 (17%)	1.78 [0.71-4.37]	0.21		
• Moderate/severe TV stenosis	37	7 (19%)	93	12 (13%)	1.58 [0.54-4.31]	0.38		
• Small/discontinuous PA	37	11 (38%)	93	9 (10%)	4.50 [1.65- 12.77]	0.004		
Preoperative ventilation	37	19 (51%)	93	30 (32%)	2.22 [1.02-4.86]	0.045	1.02 [0.26-3.71]	0.98
Preoperative inotropic support	37	8 (22%)	93	10 (11%)	2.23 [0.78-6.22]	0.125	2.68 [0.54-14.38]	0.23
Surgery ≤ 14 days	37	27 (73%)	93	43 (46%)	3.14 [1.40-7.50]	0.007	2.22 [0.67-7.89]	0.20
CPB time (min)	35	147 (88-445)	92	118 (63-365)	1.10 [1.04-1.18]*	0.002	1.11 [1.00-1.25]	0.057
Cross clamp time (min)	35	65 (12-179)	92	75 (23-133)	1.00 [0.88-1.14]*	0.97	0.89 [0.71-1.11]	0.30

N, numbers of available data.

AAI, aortic arch interruption; CI, confidence interval; CPB, cardiopulmonary bypass time; HR, hazard ratio; OR, univariate odds ratio; PA, pulmonary artery; TV, truncal valve.

*Per 10-minutes increase.

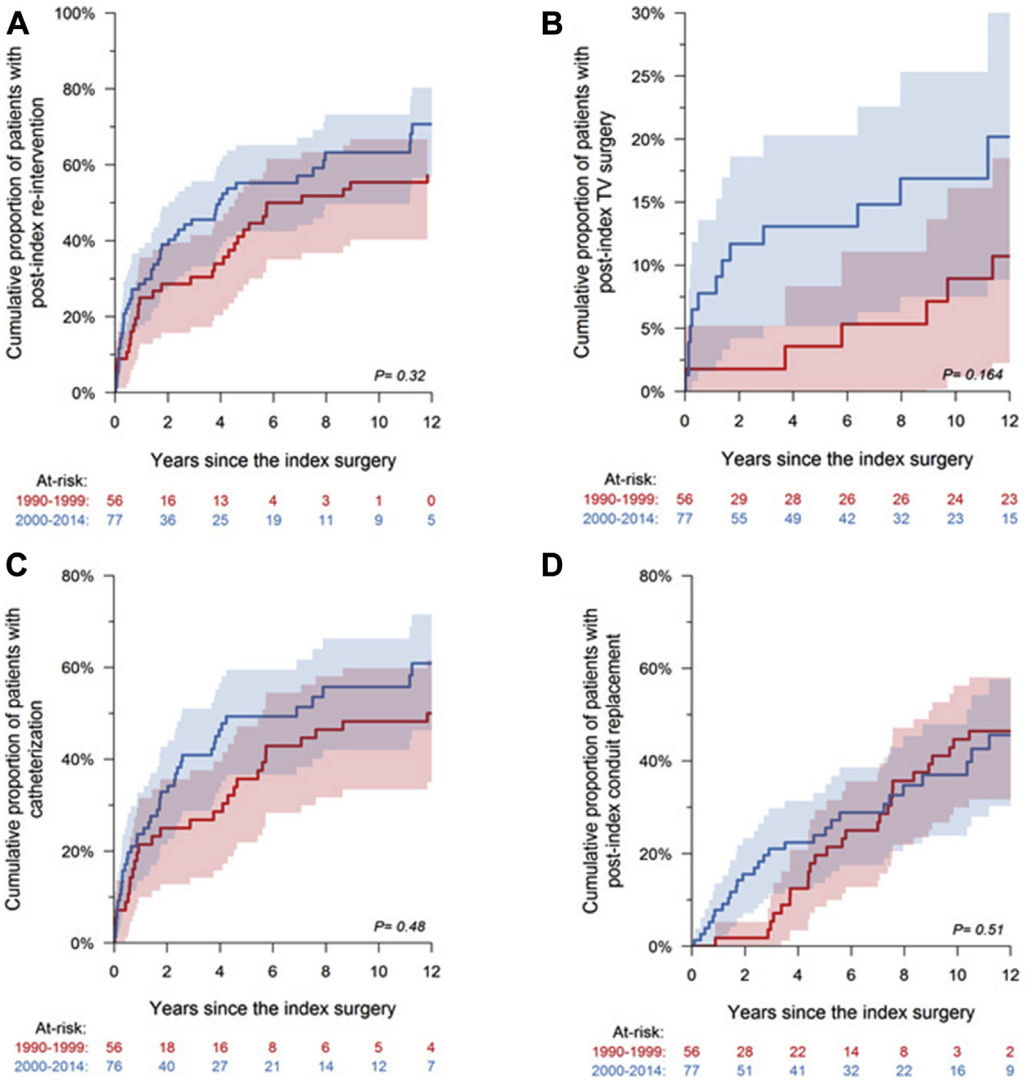

Figure 2. Cumulative proportions of reinterventions by era of (**A**) any type, (**B**) truncal valve surgery, (**C**) transcatheter conduit/PA dilation or stenting, and (**D**) conduit surgery in patients after primary biventricular repair. **Colored areas** represent 95% confidence bands.

Discussion

Current understanding of the prognosis of CAT is largely founded on studies evaluating surgical results. When compared with our patient population, most of these studies had significantly lower proportions of cases with complex pathology including aortic arch interruption[2,6,12-14] and moderate to severe TV regurgitation.[2-4,12] Yet, as Freedom prominently stated 2 decades ago,[15] "to really understand outcome, we must define the outcome for an entire cohort of patients, not just the numerator, namely those undergoing the specific procedure" as "the numerator addresses only the tip of the iceberg." This inclusive approach is particularly important for counselling and management decisions for patients affected by complex disorders such as CAT.

Prenatal outcomes

Presumably owing to the relative rarity of the disease (9 of 100,000 live births)[16] and a low rate of prenatal detection, earlier reports on fetal CAT are scarce.[7,8] Nonetheless, the prenatal rate of referral with this entity has substantially improved in our catchment area over the 25 years of our review, to 63% during the final 5 study years. Although it is unsatisfactory that a cardiac condition in which only 1 ventricular outflow is present is still frequently missed by prenatal ultrasound screening, we may underestimate the prenatal detection rate of CAT in our population. As such, families may have received diagnostic information from other health care providers and then have elected to terminate the pregnancy without a referral to us. Indeed, there are important associations with CAT and other disorders that often add to the burden these children are facing throughout life.[17,18] Genetic abnormalities were diagnosed in 40%: most commonly, microdeletion 22q11. Elective termination was the outcome of 22% of pregnancies, but this rate increased to 44% when CAT was diagnosed before 24 weeks' gestation. As with the study by Swanson et al.,[8] there was only a low (2%) rate of spontaneous intrauterine demise. Finally, a fetal diagnosis was not associated with improved postnatal survival but rather with younger gestational ages at birth and increased

odds of preoperative death. There is also increasing evidence about the added morbidity associated with a premature and early-term delivery (born before 39 weeks)[19] compared with a delivery at full term in the literature, such as increased risk of sepsis, temperature instability, hypoglycemia, and longer-term respiratory morbidity.[19-21] Considering the low risk of spontaneous in-utero death, most fetuses with CAT will not benefit from earlier induction of labour or delivery.

Pre- and postoperative outcomes

Several centres have shown that, with few exceptions, complete surgical repair during the neonatal period is feasible; even so, mortality has remained higher than for most forms of CHD. This includes data from the Society of Thoracic Surgeons Congenital Heart Surgery Database on multi-institutional outcomes of 10 benchmark operations, using operative mortality as study endpoint.[22] Among 23 participants with $\geq$ 10 surgeries from 2011 to 2014, the operative mortality of a CAT repair was 8% (0 to 27), matching the second highest mortality of all benchmark surgeries. This finding is comparable to the 7% operative mortality rate at SickKids from 2000 to 2014, and represents a significant improvement from most of the previous experiences.[3-6,12,13,22-24] The reduction in operative death in Toronto coincided with an average decrease in cardiopulmonary bypass times by 30 minutes when compared with surgeries before 2000.

Although these observations are likely a testimony of improved surgical skills and techniques, CAT is a multifaceted disease, and survival is determined by various factors such as complexity of disease. In the current series, the operative and in-hospital mortality of a simple CAT since 2000 was 2.7% and 5%, respectively. No deaths occurred beyond hospital discharge, illustrating an excellent intermediate outcome with a milder form of disease. The prognosis is, however, far less favourable when the cardiac anatomy is not "simple." Preoperative death, the outcome of one tenth of all live births with CAT, was significantly associated with significant TV stenosis, TV regurgitation, or both, and in all but 1 case, the PAs were normal sized. Williams et al.[23] also found that patients who died before surgery were significantly more likely to have moderate to severe TV stenosis than the remaining patients, although regurgitation was not a factor. In the situation of a single outlet with a significantly dysfunctional semilunar valve, the capacity of the pressure/volume-loaded heart to uphold an adequate output is limited. In the neonatal circulation, it then may become inadequate once the pulmonary vascular resistance drops and pulmonary blood flow increases, contributing to systemic and coronary hypoperfusion, ischemic organ damage, and, ultimately, to the death of the child. For cases with significant TV dysfunction, an earliest neonatal complete repair, including of the TV, should be considered to prevent this cascade of events.

Postoperative mortality was independently associated with a CAT diagnosis during study era 1, significant other cardiac lesions, and lower birth weights. Significant pathology of the TV, aortic arch, and/or branch PAs was present in 50% of study cases. The postoperative mortality of cases with complex CAT was very high (61% to 1 year) during era 1 and—despite significant improvements—continued to be substantial thereafter (25% to 1 year). Deep hypothermic circulatory arrest (DHCA) was used to provide cerebral protection for most of the aortic arch repairs. A patient with complex CAT was 4.6 times more likely to die within 30 days of surgery. As expected, reduced survival (50%) was also observed for 4 cases that were not suitable for a biventricular repair.

Reoperations

Apart from univentricular lesions, there are probably no other cardiac conditions with a similarly high prevalence of reoperations and cardiac catheterizations during childhood. As with the study by Lund et al.,[25] reinterventions for obstructed RV-PA conduits and branch PAs were particularly common, beginning soon after the initial repair and ultimately sparing less than 20% of patients during childhood.

The rate of conduit replacement or augmentation was significantly faster if the first surgery had occurred during era 1 and the child was reintervention-free at 1 year of life (Supplemental Fig. S1). We mainly used homografts in era 1 and Contegra bovine grafts in era 2, with similar diameters. Slower reoperation rates with Contegra grafts, suggestive of improved longevity, were also observed by others.[26]

Eighteen percent of our cases underwent TV surgery at or after the first surgery. This outcome was significantly associated with the presence of more than mild TV regurgitation/stenosis (19 of 36 vs no to mild, 5 of 94; OR 19.89; 95% CI, 6.53-60.59; $P < 0.001$) already at the first echocardiogram.

Limitations

The primary limitations of this study are the retrospective study design and the extended enrollment period. The functional patient status could not be objectively determined from patient charts and are therefore not reported. Echocardiograms were not systematically reviewed unless there were inconsistencies among reported findings or data were missing.

Conclusions

Our study illustrates significant improvements in prenatal detection and survival of infants with CAT. Unless there are major associated cardiac anomalies, repair of CAT in the neonatal period can be performed with excellent survival in patients. Reoperations and catheter interventions are eventualities for most patients during childhood.

Disclosures

The authors have no conflicts of interest to disclose.

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Supplementary Material

To access the supplementary material accompanying this article, visit the online version of the *Canadian Journal of Cardiology* at www.onlinecjc.ca and at https://doi.org/10.1016/j.cjca.2018.12.006.