Long-Term Survival of Patients With Coarctation Repaired During Infancy (from the Pediatric Cardiac Care Consortium)

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Patients who undergo coarctation repair during infancy have excellent early survival but long-term survival is unknown. We aimed to describe the long-term survival of patients with coarctation repaired during infancy and determine predictors of mortality. We performed a retrospective cohort study using data from the Pediatric Cardiac Care Consortium for patients with coarctation who underwent surgical repair before 12 months of age between 1982 and 2003. Long-term transplant-free survival was obtained by linkage with the National Death Index and the Organ Sharing Procurement Network. Kaplan Meier survival plots were constructed, and univariate and multivariable analyses were performed to determine predictors of mortality. We identified 2,424 coarctation patients who met inclusion criteria. At 20 years postoperatively, 94.5% of all patients and 95.8% of those discharged after initial operation remained alive, respectively. Significant multivariable predictors of mortality included surgical weight <2.5 kg (hazard ratio [HR] 3.70, 95% confidence interval [CI] 2.19 to 6.24), presence of a genetic syndrome (HR 2.40, 95% CI 1.13 to 5.10), and repair before 1990 (HR 1.91, 95% CI 1.09 to 3.34). None of the other factors examined including age at repair, gender, coarctation type, or surgical approach were found to be statistically significant. Over half of the deaths were due to the underlying congenital heart disease or other cardiovascular etiology. Overall long-term survival of patients who undergo coarctation repair during infancy is excellent. However, patients do experience small continued survival attrition throughout early adulthood. Ongoing monitoring of this cohort is necessary to assess late mortality risk. **(Am J Cardiol 2019;124:795−802)**

Coarctation of the aorta (CoA) has an estimated annual birth prevalence of 40 cases per 100,000 live births and accounts for 4% to 5% of all congenital heart disease.[1] It is commonly accompanied by other cardiac defects including aortic valve stenosis, hypoplastic aortic arch, and ventricular septal defect (VSD).[2,3] The first surgical options for CoA were developed in the middle of the twentieth century. In the past few decades, multiple surgical approaches have been used to correct this defect in infants including end-to-end anastomosis (and extended end-to-end anastomosis), subclavian flap repair, and patch angioplasty.[3] Although the early survival after CoA repair has been shown to be excellent, the long-term survival and associated risk factors are less clear, particularly among those who underwent repair in infancy. Previous studies on long-term survival after CoA repair during infancy have been single-center reports or have had relatively small numbers of infants.[4−11] In a large multicenter study from members of our research team, the overall long-term survival of patients who underwent coarctation repair was reported, but this study was not restricted to infants <1 year of age (the age at which most primary repairs are performed in the modern era) and did not analyze risk factors specific to patients with CoA.[12] Therefore, we now sought to describe the long-term survival of infants who underwent CoA repair and to determine risk factors associated with late mortality by using a large, multicenter pediatric cardiac surgical registry linked to national death and transplant records.

Methods

We performed a retrospective cohort study using data prospectively collected from the Pediatric Cardiac Care Consortium (PCCC), a large US-based registry for interventions for pediatric heart diseases that has been previously well described.[12,13] Briefly, the PCCC was established in 1982 to allow collaboration between pediatric cardiovascular centers with the aim to improve outcomes for patients with congenital or acquired heart diseases.[14] Estimates report that the PCCC contains 15% to 30% of the national volume of pediatric cardiac surgeries from 1982 to 2011.[15] This study was approved by the Institutional Review Board of Emory University.

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For this study, we included patients with CoA who had undergone initial surgical repair at <12 months of age at a participating US center from January 1982 to April 2003 (the date of the implementation of the Health Insurance Portability and Accountability Act) had a defined surgical approach recorded in their operative note, and were US residents at the time of surgery. Patients with additional cardiac lesions other than atrial septal defect, VSD, bicuspid aortic valve, aortic stenosis, or hypoplastic aortic arch were excluded from the study.

In-hospital death was defined as postoperative death during the admission for CoA repair and was recorded in the PCCC registry. Among hospital survivors, those who had adequate identifiers (first name, middle initial [if available], last name, birth day, birth month, birth year, gender, and state of birth) were previously submitted to the National Death Index (NDI) and the Organ Procurement and Transplant Network for ascertainment of vital status and transplant status complete through December 31, 2014.

A number of variables were collected and included a priori in the multivariable analyses. These included gender, age at repair, weight at repair, CoA type, surgical era, and type of CoA repair. CoA type was defined through a hierarchical approach: (1) CoA in a child with genetic syndrome as diagnosed by genetic testing or recognized clinically by phenotypic features by the center's care providers, (2) CoA with a VSD but no genetic syndrome, (3) CoA with left-sided obstruction (with accompanying hypoplastic aortic arch, bicuspid aortic valve, or aortic stenosis) but no genetic syndrome or VSD, or (4) simple CoA (with no accompanying defects other than a possible patent ductus arteriosus or atrial septal defect) without a genetic syndrome, VSD, or left-sided obstruction. Type of CoA repair was defined as end-to-end anastomosis (which included extended end-to-end anastomosis repair), subclavian flap repair, patch angioplasty, or other repair (interposition graft or end-to-side anastomosis).

Descriptive statistics were calculated for all variables of interest and included means and standard deviations or medians and ranges for continuous data and counts and percentages for categorical data. Normality of continuous variables was assessed with histograms, normal probability plots, and the Anderson-Darling test for normality. Continuous data were compared between patients who survived operative hospital admission and those who died before hospital discharge with the use of Wilcoxon rank-sum tests and comparisons between categorical variables were performed with chi-square tests, or Fisher's exact tests when the expected cell counts were <5.

To study long-term transplant-free survival overall and among those who survived to discharge after initial repair, Kaplan-Meier survival plots were constructed using all patients submitted to the NDI. Additional subplots were created to show long-term transplant-free survival by age at repair, CoA type, and type of CoA repair. Next, univariate and multivariable Cox proportional hazard regression modeling were used to determine predictors of mortality expressed as hazard ratio (HR) with associated 95% confidence interval (CI), with the variables listed mentioned previously being included in the multivariable models. In these analyses, survival without transplant after CoA repair was treated as a time-dependent outcome. Before modeling, the proportional hazard assumption was assessed using log-log survival curves and Schoenfeld residual plots, with none of the survival curves violating the proportional hazard assumption. Finally, among those who died after discharge from CoA repair, a descriptive cause of death analysis was performed using International Classification of Disease codes (ICD-9 until 1998 and ICD-10 after that year) among NDI-linked patients with general system-based etiology of death categories created based on those used in previous PCCC studies.[16] Statistical analyses were conducted using SAS 9.4 (SAS Institute, Cary, North Carolina) and statistical significance was assessed at the 0.05 level.

Results

There were 2,424 patients from 43 centers who met study inclusion criteria. In-hospital mortality occurred in 57 patients (2.4%). Of the 2,367 patients who were discharged from the hospital alive, 1,913 patients (80.8%) contained adequate identifiers to be submitted to NDI and Organ Procurement and Transplant Network. With a median follow-up of 17.7 years (twenty-fifth to seventy-fifth percentile: 14.4 to 22.0), survival among these patients was 97.5% at 1 year after repair and 95.6% at 20 years. No patients underwent transplant.

The baseline characteristics of all eligible patients are shown in Table 1. There were no differences in gender or surgical era between hospital survivors and those who died before discharge after the initial repair. Hospital survivors were more likely to be older at the time of operation (median 20 days vs 8 days), to have a greater surgical weight (3.7 kg vs 2.7 kg), to have undergone surgical repair outside of the neonatal period, and to have a simple CoA. Patients who died before hospital discharge were more likely to have CoA with VSD and the presence of a genetic syndrome. The surgical approach also differed between the 2 groups, with those who underwent patch angioplasty being more likely to suffer an in-hospital death. Baseline characteristics of patients by NDI submission status is shown in Supplemental Table 1.

The number and relative percentage of each type of CoA repair type in the PCCC is shown by era in Figure 1. Performed in 66% of the CoA repairs from 1982-1989, the subclavian flap operation was the most common type of CoA repair in the 1980s era. In the subsequent eras, the end-to-end repair replaced the subclavian flap as the dominant operation, representing 56% of the CoA repairs in the 1990's and 74% of the repairs in the early 2000's. A stable percentage of patch angioplasties occurred throughout the study period.

The overall long-term survival after CoA repair is shown in Figure 2, with survival among only those discharged after initial CoA repair shown in Figure 2. At 20 years postoperatively, 94.5% of all patients and 95.8% of those discharged after initial operation remained alive, respectively. Figure 3 shows unadjusted survival curves for CoA patients by age at repair, CoA type, and type of repair. CoA repair as a neonate was associated with worse survival than patients who underwent repair at 6 to 12 months of age, with much of the difference becoming apparent in the first few postoperative years. Patients with a genetic syndrome had worse survival compared with isolated CoA. Among

Table 1
Summary of patients overall and by in-hospital mortality after definitive repair of coarctation

Characteristics	Overall n = 2,424	Survived to hospital discharge n = 2,367	In-hospital mortality after repair n = 57	p value*
Age at repair (days) Median (25th−75th)	19 (9 − 74)	20 (9 − 75)	8 (5 − 17)	**<0.001**
Age group				**<0.001**
Neonate (<28 days)	1388 (57.3%)	1340 (56.6%)	48 (84%)	
1 month ≤ 6 months	792 (32.7%)	784 (33.1%)	8 (14%)	
6 − 12 months	244 (10.1%)	243 (10.3%)	1 (2%)	
Sex				0.36
Male	1502 (62.0%)	1470 (62.1%)	32 (56%)	
Female	922 (38.0%)	897 (37.9%)	25 (44%)	
Surgical weight (kg) Median (25th − 75th) (n = 2,407)	3.62 (3.0 − 4.6)	3.67 (3.07 − 4.64)	2.71 (2.29 − 3.28)	**<0.001**
Surgical weight < 2.5 kg	230 (9.6%)	213 (9.1%)	17 (31%)	**<0.001**
Coarctation Type				**<0.001**
Simple coarctation	1178 (48.6%)	1164 (49.2%)	14 (25 %)	
Left-sided obstruction	438 (18.1%)	433 (18.3%)	5 (9%)	
Ventricular septal defect	652 (26.9%)	625 (26.4%)	27 (47%)	
Genetic syndrome†	156 (6.4%)	145 (6.1%)	11 (19%)	
Surgical era				0.45
1982 − 1989	386 (15.9%)	375 (15.8%)	11 (19%)	
1990 − 1999	1355 (55.9%)	1321 (55.8%)	34 (60%)	
2000 − 2003	683 (28.2%)	671 (28.4%)	12 (21%)	
Type of coarctation repair				**<0.001**
End-to-end	1344 (55.5%)	1325 (56.0%)	19 (33%)	
Subclavian flap	862 (35.6%)	842 (35.6%)	20 (35%)	
Patch angioplasty	204 (8.4%)	189 (8.0%)	15 (26%)	
Interposition graft	12 (0.5%)	9 (0.4%)	3 (5%)	
End to side	2 (<0.1%)	2 (<0.1%)	0 (0%)	

* Comparison of those who survived to hospital discharge versus those who had in-hospital mortality following repair.
† Genetic syndromes included 97 with Turner syndrome, 25 with Trisomy 21, 8 with DiGeorge syndrome, 5 with Noonan syndrome, and 21 with other syndromes or chromosomal abnormalities.
Significant findings indicated in bold.

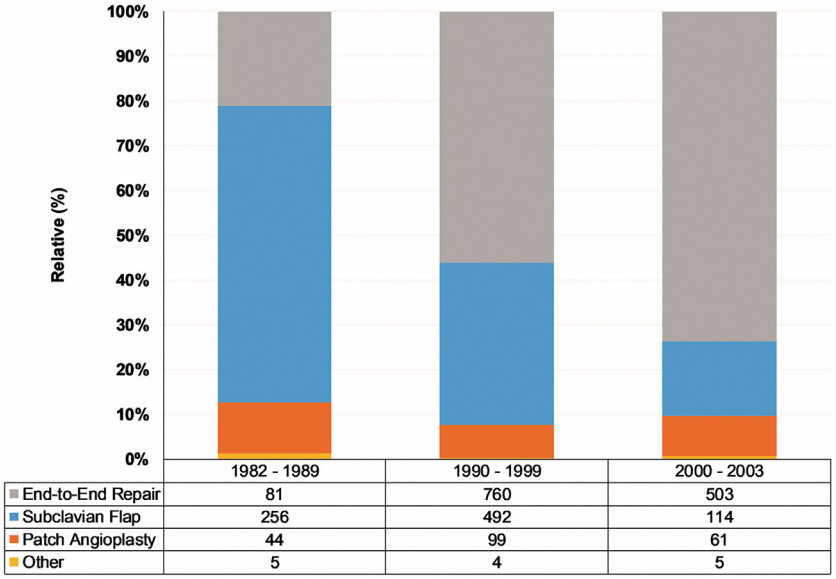


Figure 1. Surgical approach by era. Surgical strategy changed across time with subclavian flap being used primarily in the earlier years and end-to-end repair being used in the later years (p <0.001).

the other types of CoA there were no significant differences in survival. Patch angioplasty and subclavian flap repair showed decreased survival compared with patients repaired by end-to-end anastomosis, with much of the mortality for patch angioplasty coming early but mortality for subclavian flap repair coming both early and late.

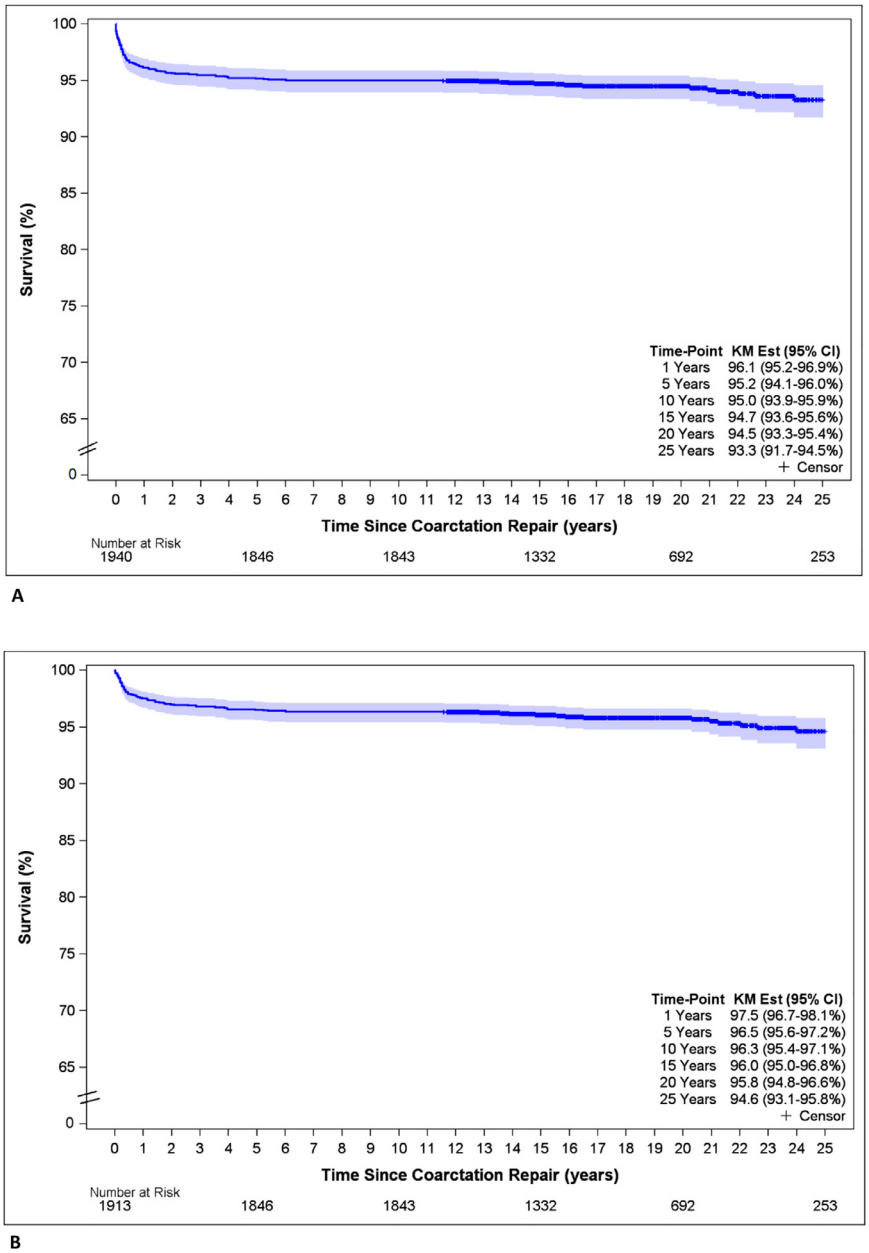


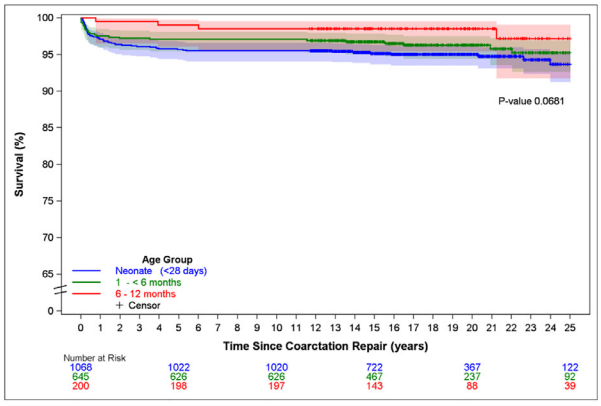
Figure 2. Overall long-term survival after coarctation repair *(A)* and survival among only those discharged after initial coarctation repair *(B)*. *Solid blue* represents the nonparametric Kaplan-Meier (KM) survival estimates. The *shaded area* is the 95% confidence interval for the KM estimates. (Color version of figure is available online.)

Univariate and multivariable analyses of predictors of mortality are shown in Table 2. In the multivariable analyses, weight of <2.5 kg at time of repair (HR 3.70 compared with weight ≥2.5 kg, 95% CI 2.19 to 6.24), presence of a genetic syndrome (HR 2.40 compared with simple CoA without a syndrome, 95% CI 1.13 to 5.10), and surgical era in the 1980s (HR of 1.91 compared with repair in the 1990s, 95% CI 1.09 to 3.34) were significantly associated with increased long-term mortality. Age at repair, gender, other CoA types, type of surgical repair, and repair in the year 2000 and beyond were not statistically significant in the multivariable analyses.

Causes of death are shown grouped by general system-based etiologies in Figure 4 and Supplementary Table 2. Over half of the deaths were due to an underlying cause of congenital heart disease or other disease of the cardiovascular system, including 1 report of stroke as an underlying cause of death (and 1 additional report of stroke as a contributing cause of death). Other substantial causes included congenital malformations/chromosomal abnormality, external causes of injury, and diseases of the respiratory system.

Discussion

In this retrospective cohort study from the PCCC, the largest study of its kind in this population to our knowledge, we found that the survival to early adulthood was excellent



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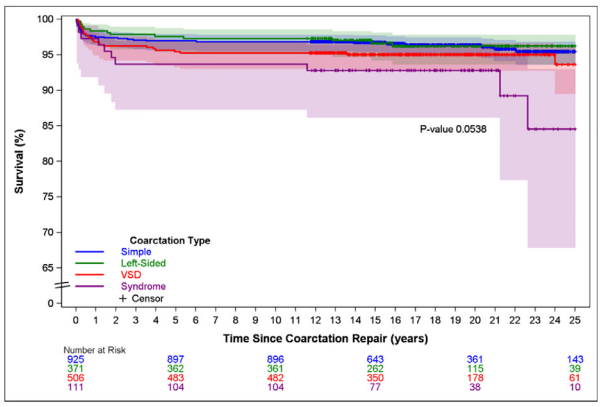

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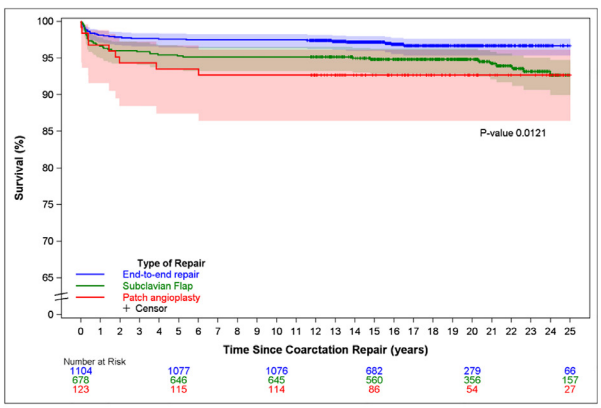

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Figure 3. Unadjusted Kaplan-Meier survival curves stratified by age *(A)*, coarctation type *(B)*, and coarctation repair type *(C)*. *(A)* Survival plots with log-rank p values demonstrate that patients operated on between 6 and 12 months of age had improved survival over neonates; *(B)*survival analysis demonstrated that patients with coarctation and genetic syndrome had worse survival compared to those with simple coarctation; *(C)*survival analysis demonstrated that patients repaired by patch angioplasty of subclavian flap have worse survival compared with patients with end-to-end repair.

among those who survive to hospital discharge after initial surgical repair of CoA during infancy. Overall, survival in this large cohort of patients followed for nearly 2 decades are similar to previously reported results.[4−10,17] However, the large number of patient-years in this cohort allowed us to identify risk factors for late mortality such as low surgical weight, earlier era of surgery, and the presence of a genetic syndrome. Although surgical repair type was a univariate predictor of late mortality, this association was no longer significant when accounting for other variables.

The majority of the patients in our study underwent an end-to-end anastomosis; however, the subclavian flap repair was more common in the earlier era. Patients who had accompanying left-sided obstructive lesions or VSDs did not appear to do worse than those with isolated CoA in the long term. Only the presence of a genetic syndrome significantly impacted survival. Past results have shown that patients with an accompanying VSD have increased early mortality.[5,18−20] A couple of small, single center reports of long-term CoA follow-up reported that VSD was associated with late mortality as well.[5,21] Yet, here we demonstrate that in those who survive operative hospital admission, survival of CoA patients with a VSD is similar to that of those with isolated CoA. Additionally, the presence of left-sided obstructive lesions has been shown in the past to impact results, as patients with a hypoplastic arch have been shown to be at higher risk operative mortality as well as for restenosis and reintervention.[19,21−23] The presence of an accompanying bicuspid valve also leads to higher valve-related reinterventions.[24] Despite this, our study suggests that CoA patients with left-sided obstructive lesions do not have lower survival rates through 20 years postoperation. Genetic syndrome was associated with a significantly decreased hazard of mortality on multivariable analysis.

Other factors that influenced the hazard of mortality on multivariable analysis included low surgical weight and era of operation. Previous reports from this era are mixed, with some suggesting surgical weight impacts both reintervention rates and mortality[25] and others finding no difference in patients with a low surgical weight.[4,21,23,26] Although the age of CoA repair was not a significant predictor of mortality on multivariable analysis, repair from 6 to 12 months of life was associated with a significantly decreased hazard of mortality on univariate modeling. Neonatal repair has been found to have higher rates of complications and reinterventions[23] but some reports show age at surgery has not been predictive of late survival.[5] Our results suggest that having a surgical weight ≥2.5 kg may be a more important factor for predicting long-term survival than the exact timing of surgery within the first 12 months of life. It is unclear why this is the case: possible explanations include that successful repair is more challenging in a smaller patient[27] or that low birth weight infants may be more likely to have comorbidities such as intraventricular hemorrhage.[28] With regards to surgical era, compared with the 1990s era, we found that patients who had operations in the 1980s had increased long-term mortality. Pillutla et al[29] found that mortality rates of patients with CoA have been consistently decreasing from 1980 to 2005. The effect of era may represent changes in the type of surgical repair, the surgical approach, or improvements in postoperative management.

The type of surgical CoA repair has been shown to impact patient morbidity and reintervention rates.[30] However, the impact of repair type on late survival is less clear with excellent reports from both subclavian flap and end-to-end anastomosis.[4,10] Our study shows that, although the end-to-end anastomosis had superior results compared with subclavian flap and patch angioplasty on univariate

Table 2
Risk factors associated with mortality following hospital discharge from coarctation repair

Risk factors	Univariate		Multivariable*	
	HR (95% CI)	p value	HR (95% CI)	p value
Age group				
Neonate (<28 days)	Reference	–	–	–
1 month ≤ 6 months	0.73 (0.45-1.16)	0.18	0.89 (0.55-1.44)	0.62
6 – 12 months	0.36 (0.13-0.98)	**0.047**	0.51 (0.18-1.43)	0.20
Female (vs. male)	1.09 (0.70-1.67)	0.71	0.83 (0.52-1.33)	0.44
Weight <2.5 kg (vs. ≥2.5)	3.86 (2.35-6.33)	**<0.0001**	3.70 (2.19-6.24)	**<0.0001**
Coarctation type				
Simple coarctation	Reference	–	–	–
Left-sided obstruction	0.92 (0.49-1.74)	0.80	1.03 (0.54-1.96)	0.93
Ventricular septal defect	1.35 (0.82-2.24)	0.24	1.28 (0.76-2.14)	0.36
Genetic syndrome	2.40 (1.19-4.84)	**0.01**	2.40 (1.13-5.10)	**0.02**
Surgical era				
1982 – 1989	1.69 (1.00-2.83)	0.052	1.91 (1.09-3.34)	**0.02**
1990 – 1999	Reference	–	–	–
2000 – 2003	0.85 (0.48-1.50)	0.57	0.98 (0.54-1.75)	0.93
Type of coarctation repair				
End-to-end repair	Reference	–	–	–
Subclavian flap	1.83 (1.16-2.88)	**0.03**	1.41 (0.86-2.30)	0.17
Patch angioplasty	2.26 (1.08-4.72)	**0.01**	1.73 (0.81-3.66)	0.16

* Each analysis is adjusted for the other factors in the table.
Significant findings indicated in bold.

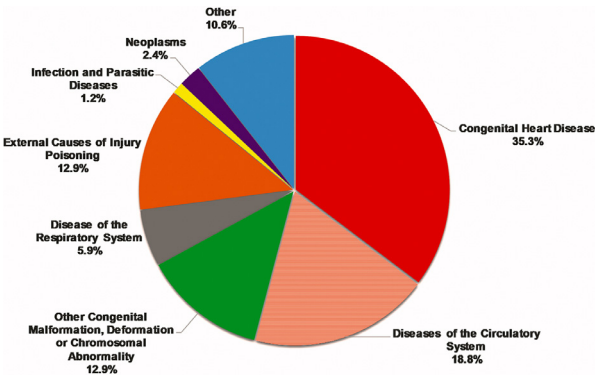

Figure 4. Underlying cause of death among patients with a history of surgery for coarctation of the aorta during infancy.

analysis, there was not a significant late survival difference between the different types of surgery when accounting for other factors in the multivariable analyses. It is possible that the era effect is the key factor; that is, given that most subclavian flap repairs occurred in the 1980s and surgeries in the 1980s had worse long-term outcomes compared with later years, the era in which 1 had surgery may be more important than the type of surgery. Because the number of deaths is relatively low to fully evaluate this hypothesis at this time, further follow-up is required to address this question in the future.

The patients who suffered late deaths in our study most commonly died from the underlying congenital heart disease and other cardiovascular etiologies. CoA is known to be associated with persistent endovascular abnormalities. Early changes in vascular gene expression in the proximal aorta may impact the development of hypertension.[31] Patients with repaired CoA additionally have abnormal left ventricular diastolic function and aortic stiffness, even when early CoA repair is achieved.[11,32] Patients are also at risk of recoarctation, aortic aneurysm formation, endocarditis, and cerebrovascular hemorrhage, all of which likely contribute to late causes of death.[3,33] Thus, CoA needs to be regarded as a lifelong disease that necessitates long-term surveillance.

Our study has several limitations. First, our study is an observational study with the limitations inherent of retrospective studies. Second, the PCCC has insufficient data regarding other variables that may be important to long-term survival. These include race/ethnicity, socioeconomic status, and preoperative clinical status. Third, PCCC lacks information regarding severity of lesions. Thus, we are unable to comment on the severity of initial coarctation or any associated lesions. It is possible that many patients in this study with bicuspid aortic valve had no significant obstruction, but we chose to treat all bicuspid aortic valve patients as part of the left-sided obstructive lesions group for this study. Fourth, although every effort was made to link patients with the NDI, we cannot exclude potential selection bias given that a small portion of patients could not be successfully linked or that the NDI may have missed certain deaths. Although the NDI linkage does have a high sensitivity for identifying known deaths, it is certainly not perfect, particularly for identifying cause of death.[15] Finally, the PCCC has limited data on long-term complications, with information being available only if the patient returned to a center participating in the PCCC at the time of the complication and received an intervention for the complication. This prevents analysis of CoA restenosis, reintervention rates, cerebrovascular events, and other complications that may vary, particularly by type of surgical repair.

Long-term survival of patients with CoA repaired in infancy is excellent, but there is a small ongoing risk of

mortality into early adulthood, primarily due to an underlying cause of congenital heart disease or other disease of the cardiovascular system. Patients at higher risk included those with surgical weight <2.5 kg, those with genetic defects, and those who underwent repair in an earlier era. The type of surgical approach was not associated with significant differences in survival. Continued surveillance of adult CoA survivors will help to direct future management approaches and guide adult surveillance protocols.

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Disclosures

The data reported here have been supplied by UNOS as the contractor for the Organ Procurement and Transplantation Network (OPTN). The interpretation and reporting of these data are the responsibility of the author(s) and in no way should be seen as an official policy of or interpretation by the OPTN or the US Government.

Supplementary materials

Supplementary material associated with this article can be found in the online version at https://doi.org/10.1016/j.amjcard.2019.05.047.

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