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Congenital Disease

Coarctation of the Aorta

Lifelong Surveillance Is Mandatory Following Surgical Repair

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Objectives The objective of our study was to review the long-term outcomes of patients undergoing surgical repair of aortic coarctation.

Background Surgical repair of aortic coarctation has been performed at the Mayo Clinic, Rochester, Minnesota, for over 60 years.

Methods Between 1946 and 2005, 819 patients with isolated coarctation of the aorta underwent primary operative repair. Medical records were reviewed and questionnaires mailed to the patients.

Results Mean age at repair was 17.2 ± 13.6 years. The majority (83%) had pre-operative hypertension. Operations included simple and extended end-to-end anastomosis (n = 632), patch angioplasty (n = 72), interposition grafting (n = 49), bypass grafting (n = 30), and subclavian flap or "other" (n = 35). Overall early mortality (<30 days) was 2.4%. In the previous 30 years (n = 225), there were no operative deaths. Mean follow-up was 17.4 ± 13.9 years, with a maximum of 59.3 years. Actuarial survival rates were 93.3%, 86.4%, and 73.5% at 10, 20, and 30 years, respectively. When compared to an age- and sex-matched population, long-term survival was decreased ($p < 0.001$). Older age at repair (>20 yrs) and pre-operative hypertension were associated with decreased survival ($p < 0.001$). Patients age <9 years age at repair had significantly less hypertension at 5 to 15 years of follow-up ($p < 0.001$). Rates of freedom from re-intervention on the descending aorta were 96.7%, 92.2%, and 89.4% at 10, 20, and 30 years, respectively. Younger age at time of repair ($p < 0.001$) and an end-to-end anastomosis technique ($p < 0.001$) were independently associated with lower rates of re-intervention on the descending aorta.

Conclusions Primary repair of isolated coarctation of the aorta was performed with a low rate of mortality. However, long-term survival was reduced compared with that in an age- and sex-matched population, and many patients required further reoperation. These findings emphasize that patients with aortic coarctation need early recognition and intervention, as well as lifelong informed follow-up. (J Am Coll Cardiol 2013;62:1020–5) © 2013 by the American College of Cardiology Foundation

Aortic coarctation is defined as a narrowed aortic segment, most commonly located near the ligamentum arteriosum adjacent to the left subclavian artery (1). It can be discrete, long, or complex, involving the aortic arch or isthmus, and may have collateral vessels (1). Coarctation of the aorta has an estimated incidence of 1 in 2,500 live births, with a 2:1 predominance in males (2,3). In historical series, untreated aortic coarctation led to a mortality of more than 80% by age 50 years due to complications including aortic rupture, heart failure, and intracranial hemorrhage (4).

The first successful repair of aortic coarctation was described by Dr. Crafoord in 1945 (5), and the first repair at the Mayo Clinic was in 1946. Following surgical repair of aortic coarctation, patients experience improved survival, with age at time of repair being the most important predictor of survival (6). In general, significant coarctation repair should occur as early as possible. However, the optimal timing of this intervention is still not clear, as it is a balance between the risks for early and late mortality, reoperation, and the long-term effects of hypertension.

Our objective was to review the experience of repair of aortic coarctation over 60 years at the Mayo Clinic to identify predictors of long-term survival, reoperation, and late hypertension.

Methods

Institutional review board approval was obtained, which obviated individual patient consent. From August 1946 to

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July 2005, 819 patients had primary operative repair of isolated aortic coarctation. We excluded patients who had complex left-sided cardiac lesions, interrupted aortic arch, or Shone's complex.

Perioperative data were collected using all available medical records. Follow-up data were collected using all medical records, and surveys were mailed to all patients not known to be deceased. Survival was ascertained through Accurint.com.

As a part of quality assurance at the Mayo Clinic, all patients who have undergone cardiac surgery are sent a survey on a scheduled basis. This survey includes a question that asks whether a diagnosis of high blood pressure was present, as well as inquiring regarding current medications and any cardiac catheterization procedures or surgical procedures performed. In patients in whom an intervention is reported, medical records are requested from outside institutions in an attempt to confirm the procedure indication and type. The presence of hypertension was determined through repeated elevated blood pressure measurements using standard definitions of hypertension (adults: systolic blood pressure ≥140 mm Hg and diastolic blood pressure ≥140 mm Hg) (7) and for children as systolic and/or diastolic blood pressure in the ≥95th percentile (8), the prescription or report of antihypertensive medications, or the self-report of a diagnosis of hypertension.

Statistical analyses were performed using SAS version 9.1 (SAS Institute Inc, Cary, North Carolina). Data are presented as mean ± SD, median (range), or number (%), as appropriate. Univariate and stepwise multivariate models were created using Cox proportional hazards. Kaplan-Meier curves were compared using a log-rank test. Cutoff values were identified using an SAS macro designed to find the best cutoff point of a continuous variable based on a chi-square statistic for a binomial outcome.

Results

Pre-operative characteristics are listed in Table 1. Operations included end-to-end anastomosis in 631 patients, patch angioplasty in 73, interposition graft in 49, bypass graft in 30, and subclavian flap or other in 36. These operations were predominantly performed prior to the 1980s (n = 612; 74.7%). The type of repair has changed over the decades, with older patients and more complex repairs predominating in later decades (Fig. 1). Overall early mortality (<30 days) was 2.4%. In the previous 30 years (n = 225), there were no operative deaths.

Mean follow-up was 17.4 ± 13.9 years, with a maximum of 59.3 years. Four hundred fifty-nine patients had follow-up >30 days, and an additional 122 surveys were returned. Thus, data on mid- or late-term follow-up was obtained from 70.8% of all patients.

Actuarial survival rates were 93.3%, 86.4%, and 73.5% at 10, 20, and 30 years, respectively. Mean age of death was 34.2 ± 20.1 years. When compared to an age- and sex-matched population, long-term survival was decreased ($p < 0.001$) (Fig. 2). Older age at repair (>20 years) and pre-operative hypertension were associated with decreased survival ($p < 0.001$). On multivariate analysis, only increasing age was associated with mortality (hazard ratio [HR]: 1.06; 95% CI: 1.04 to 1.07; $p < 0.001$) (Fig. 3).

Abbreviations and Acronyms

ASD = atrial septal defect
NYHA = New York Heart Association
PFO = patent foramen ovale

Rates of freedom from re-intervention on the descending aorta were 96.7%, 92.2%, and 89.4% at 10, 20, and 30 years, respectively (Fig. 4). Older age (HR: 0.93; 95% CI: 0.91 to 0.96; $p < 0.001$) and an end-to-end anastomosis technique (HR: 0.11; 95% CI: 0.07 to 20; $p < 0.001$) were independently associated with lower re-intervention rates on the descending aorta. Patients who had initial repair before age 1 year (n = 116), had a 31.1% re-intervention rate on the descending aorta rate at 30 years' follow-up. The highest rate of re-intervention on the descending aorta was in patients who were <5 years of age at the time of operative repair (73.3%; 95% CI: 64.1% to 83.9% at 30 years; $p < 0.001$; n = 192). In patients <5 years of age, simple and extended end-to-end anastomosis technique was associated with a decreased re-intervention rate, $p < 0.013$.

Rates of freedom from any cardiac reoperation were 92.8%, 85.7%, and 76.6% at 10, 20, and 30 years, respectively. The majority of reoperations were for repair of recoarctation or aortic valve disease (Table 2).

Data on follow-up regarding blood pressure status were available from 551 patients (67.3%). Mean follow-up was 17.4 ± 13.8 years, with a maximum of 59.3 years. The percentages of patients with hypertension at various follow-up intervals are presented in Fig. 5. The only predictor of hypertension up to 5 years postoperatively was older age at the time of operation ($p < 0.001$). A cutoff value of 9 years was identified; patients 9 years of age or older at the age of repair had significantly more hypertension at 5 to 15 years of follow-up (relative risk: 4.1; 95% CI: 2.1 to 8.4; $p < 0.001$).

Table 1 Pre-Operative Characteristics (N = 819)

Characteristic	Value
Age at operation	
Mean ± SD, yrs	17.2 ± 13.6
Range	1 day to 72.2 yrs
Age group	
≤1 yr	116
>1–≤5 yrs	76
>5–≤10 yrs	123
>10–≤20 yrs	235
>20 yrs	269
Female	243 (30)
Pre-operation hypertension	683 (83)
NYHA class III or IV	32 (5)

Values are mean ± SD, n, or n (%).
NYHA = New York Heart Association.