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## Surgical repair of aortic coarctation in adults: half a century of a single centre clinical experience

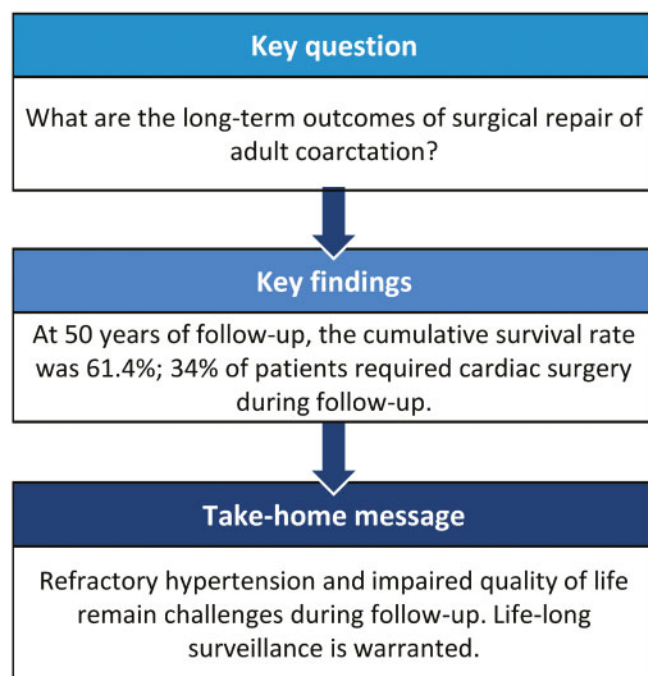
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### Abstract

**OBJECTIVES:** Coarctation of the aorta (CoA) is rarely diagnosed and treated in adults and nowadays mostly treated with percutaneous techniques. The objective of this study is to report the long-term outcomes and health-related quality of life in a unique cohort of patients treated with an open surgical repair for their primary adult CoA.

**METHODS:** Ninety adult patients underwent primary surgical repair of CoA at our tertiary referral centre between 1961 and 2008 when the treatment strategy for adult CoA was exclusively surgical.

**RESULTS:** The median age at surgery was 24 years (interquartile range 20–36 years), and 39 patients (43%) were asymptomatic at presentation. CoA was located paraductally in most patients (64%), and bicuspid aortic valve was present in 39 (43%) patients. Surgical reconstruction of CoA with an end-to-end anastomosis was performed in majority of the patients (57%). Overall, in-hospital mortality occurred in 1

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patient (1%). There was no in-hospital stroke, spinal cord ischaemia, renal replacement therapy or respiratory failure. The cumulative survival was 97.7%, 89.5%, 82.6%, 70.9% and 61.4% at 10, 20, 30, 40 and 50 years, respectively. Thirty-one patients (34%) required an additional cardiac surgery during follow-up. The majority of patients (77%) suffered from refractory hypertension even after decades of surgery. Compared with the matched Dutch population, patients reported a lower social functioning, mental health, vitality and general health with a higher body pain.

**CONCLUSIONS:** Patients with native adult CoA have low in-hospital morbidity and mortality when treated with an open surgical reconstruction. However, refractory hypertension and impaired quality of life remain important challenges during follow-up.

**Keywords:** Coarctation • Adult • Surgical repair • Health-related quality of life

#### ABBREVIATIONS

BAV	Bicuspid aortic valve
CoA	Coarctation of the aorta
CT	Computed tomography
IQR	Interquartile range
MC	Medical Center
MRI	Magnetic resonance imaging
QoL	Quality of life
SF-36	Short Form 36-Item Health Survey

## INTRODUCTION

Coarctation of the aorta (CoA) is a congenital anomaly, most often diagnosed in neonates and infants due to symptomatic presentation. It accounts for 5–8% of all congenital heart defects and the estimated prevalence is 3 per 10 000 live births [1]. However, adolescents as well as adults can also be diagnosed with primary CoA of the native aorta as first-time diagnosis [2]. In this untreated and underdiagnosed older age group, primary adult CoA can for example be recognized as an incidental finding by a primary care physician who notices hypertension, heart murmur or decreased femoral pulses as part of a routine annual physical examination. Untreated patients may suffer from systemic arterial hypertension, heart failure, aortic aneurysm, aortic dissection and are at risk for early mortality, usually in the third or fourth decade [3].

A wide spectrum of concomitant cardiovascular anomalies is associated with CoA. Bicuspid aortic valve (BAV) is most frequently involved, with a reported range of 40–80% [1, 2]. Hence, aortic coarctation should be considered as a complex composite of cardiovascular disorders. Historically, surgical intervention for patients with CoA results in good anatomical reconstruction, with acceptable long-term survival, but late complications are not unusual so these patients require monitoring with imaging [1, 4, 5].

Although surgery is the primary treatment modality in children, endovascular treatment of coarctation with balloon angioplasty and stenting is used increasingly in adults [6]. Although surgical repair is silently being replaced by interventional treatment, an explicit presentation of long-term surgical results can be of help to put treatment alternatives into a realistic perspective. Furthermore, the current literature comprises mainly paediatric patient population. Hence, reports on adult coarctation presenting during adult life most frequently represent cases of re-coarctation, following previous transcatheter or surgical therapy. To date, no published data exists on adult patients with isolated primary CoA with long-term follow-up.

In this regard, the aim of the present study was to report on our unique consecutive clinical experience over a period of more than 5 decades treating patients with primary adult aortic coarctations with an open surgical repair at the Erasmus University Medical Center (MC) in Rotterdam, the Netherlands and to provide data allowing benchmarking for percutaneous treatment.

## METHODS

### Study design

Between July 1961 and October 2008, 90 consecutive adult patients underwent isolated primary open repair of CoA at the Erasmus MC in Rotterdam, the Netherlands. The patients were recruited between this period, because until 2008, in our centre adult CoA was exclusively treated with surgery, while thereafter the predominant strategy for this issue was percutaneous intervention. All patients older than 16 years of age with 'isolated CoA', defined as having no associated congenital heart disease except for BAV, persistent ductus arteriosus or small atrial or ventricular septal defects, were included.

The medical ethics commission reviewed and approved this study prior to its onset and waived individual informed consent (MEC-15-581). The clinical charts of all patients were retrospectively reviewed and all clinically gathered data including the occurrence of events during follow-up and the cause of death was registered and reported according to the EACTS/ESCVS best practice guidelines for reporting treatment results in the thoracic aorta [7]. Data were collected from hospital records, operative reports and correspondence with the patients.

Currently, magnetic resonance imaging (MRI) or computed tomography (CT) angiography with 3-dimensional reconstruction is the most adequate test to image the precise location and anatomy of the coarctation and the entire aorta, as well as collateral vessels.

### Clinical follow-up

The annual follow-up was performed as a regularly scheduled outpatient visit with blood pressure monitoring and CT or MRI. The national civil registry was consulted to confirm the alive status and these patients were contacted by mail for quality of life (QoL) and long-term follow-up. The follow-up was complete in 90% of all patients; 9 patients were lost to follow-up due to migration. Operative records of 4 patients could not be retrieved.

Operative events were defined as those occurring within 30 days after surgery or any event later during the same postoperative hospital stay. Hypertension was defined as a blood pressure of >140/90 mmHg on 2 separate occasions at rest or the

need for antihypertensive medications. Re-coarctation was defined as having hypertension and a resting blood pressure gradient  $\geq 20$  mmHg between the right arm and leg [1]. Re-coarctation was considered significant if it required percutaneous or surgical repair.

## Operative technique

Over the study period different techniques for the open repair of CoA's were used. Over time, the choice of a certain surgical technique depended on the patient's characteristics, e.g. the length of the coarctation segment, extent of collaterals and the surgeon's preference. The majority of patients underwent direct end-to-end repair. Other commonly used techniques were extended end-to-end anastomosis, patch aortoplasty and graft interposition. Most frequently the traditional clamp-and-sew technique was used without distal aortic perfusion or spinal neuromonitoring.

The patient was placed in the right lateral decubitus position and a left thoracotomy incision was made in the fifth intercostal space for exposure of the proximal descending thoracic aorta. After opening the mediastinal pleura, prominent intercostal collaterals arising from the aorta were identified and ligated if considered necessary. When test clamping of the aorta showed no or limited fall of the blood pressure (femoral mean pressure), the decision was made to continue without the use of the extracorporeal circulation. In case the distal aortic blood pressure decreased significantly and fell below a mean of 50 mmHg with loss of systolic augmentation during test clamping, left heart bypass was initiated. Distal aortic perfusion was established with pump-assisted bypass with direct cannulation of the aortic arch and the descending thoracic aorta.

## Quality of life

The quality-of-life assessment was performed using the Short Form 36-Item Health Survey (SF-36) [8]. The questionnaire incorporates 36 scale-rated health-related questions, arranged into 8 multi-item sections that are not disease-specific. These questions measure functioning in different aspects of daily life: 'physical functioning'; physical health related to age- and role-specific activities: 'role physical', 'bodily pain', 'general health', 'vitality', 'social functioning'; personal feelings of performance in age- and role-specific activities: 'role emotional' and 'mental health'. Questions used in the SF-36 form do not distinguish between body pain caused by wound scar or otherwise. All surviving patients received the SF-36 (Dutch version) questionnaire by mail and were asked to return the completed questionnaire. To compare these outcomes with the general age-matched Dutch population, Dutch norms that were previously published by Aaronson et al. [9] were used.

## Statistical analysis

Normality of continuous variables was tested with the Shapiro-Wilk and Kolmogorov-Smirnov tests. Continuous data were presented as mean  $\pm$  standard deviation or median and interquartile range (IQR), as appropriate. Categorical data are presented as proportions. Cumulative survival and freedom from reintervention were analysed using the Kaplan-Meier method. Censoring

**Table 1:** Baseline characteristics

Characteristics	Entire cohort (N = 90)
Operation age (years), median (IQR)	24 (20–36)
Male gender, n (%)	53 (59)
Asymptomatic, n (%)	39 (43)
Symptomatic, n (%)	
Claudication/limb pain	20 (22)
Chest pain	12 (13)
Palpitations	12 (13)
Dyspnoea	23 (26)
Fatigue	28 (31)
Headache	16 (18)
Back pain	1 (1)
Vertigo/dizziness	16 (18)
Preoperative SBP (mmHg), median (IQR)	175 (157–201)
Preoperative DBP (mmHg), median (IQR)	96 (85–110)
Hypertension, n (%)	90 (100)
Antihypertensive treatment, n (%)	40 (44)
Femoral pulsations, n (%)	
Good	11 (12)
Weak	60 (67)
Absent	19 (21)
COPD, n (%)	0 (0)
History of CVA, n (%)	1 (1)
Diabetes, n (%)	0 (0)
Renal dysfunction, n (%)	0 (0)
Coronary artery disease, n (%)	1 (1)
LV function, n (%)	
Normal (LVEF $\geq 50\%$ )	80 (89)
Moderately reduced (LVEF 35–49%)	10 (11)
Reduced (LVEF $< 35\%$ )	0 (0)
LV morphology, n (%)	
Normal	36 (40)
Hypertrophic	41 (46)
Dilated	5 (6)
Dilated + hypertrophic	8 (9)
Associated anomalies, n (%)	
BAV	39 (43)
VSD	5 (6)
ASD	1 (1)
PDA	4 (4)
Hypoplastic isthmus	7 (8)
Hypoplastic aortic arch	0 (0)
Poststenotic dilatation	19 (21)
Aortic regurgitation	21 (23)
Aortic stenosis	8 (9)
Combined aortic valve disease	5 (6)
Turner syndrome	3 (3)
Previous cardiac operation, n (%)	10 (11)

ASD: atrial septal defect; BAV: bicuspid aortic valve; COPD: chronic obstructive pulmonary disease; CVA: cerebrovascular accident; DBP: diastolic blood pressure; IQR: interquartile range; LV: left ventricle; LVEF: left ventricular ejection fraction; PDA: patent ductus arteriosus; SBP: systolic blood pressure; VSD: ventricle septal defect.

of patients occurred at the time of their last follow-up. Age- and gender-matched survival in the general population was calculated using the Dutch population life tables (<http://statline.cbs.nl/>). Self-reported QoL comparison of the study population with the general age-matched Dutch population was performed using the Wilcoxon rank-sum test with Bonferroni correction. This latter correction indicates that the allowable significance level for each SF-36 subscale was  $P$ -value  $< 0.00625$  (0.05/8 subscales). A  $P$ -value  $< 0.05$  was considered significant throughout the analyses. Statistical analyses were performed with IBM SPSS Statistics

**Table 2:** Procedure characteristics

Characteristics	Entire cohort (N = 90)
Location of coarctation, n (%)	
Preductal	7 (8)
Juxtaductal	58 (64)
Postductal	25 (28)
Surgical technique, n (%)	n/N = 86
End-to-end anastomosis	49 (57)
Extended end-to-end anastomosis	11 (13)
Subclavian flap	0 (0)
Patch aortoplasty	11 (13)
Graft interposition	15 (17)
Left heart bypass	9 (11)

for Mac, version 23 (IBM Corp., Armonk, NY, USA) and R statistical program (R Foundation for Statistical Computing, Vienna, Austria. URL <https://www.R-project.org/>).

## RESULTS

During the study period, 90 adult patients underwent open surgical repair of primary CoA. The median age at surgery was 24 years (IQR 20–36 years) and 53 (59%) were male (Table 1). Thirty-nine (43%) patients were asymptomatic and the CoA was incidentally discovered during workup for cardiac murmur or military medical examination by measuring hypertension. Among 51 (57%) symptomatic patients, the most common symptom was fatigue in 28 patients (31%), followed by shortness of breath in 23 (26%) and leg claudication in 20 (22%). Upper body hypertension was present in all patients (100%) and 40 (44%) patients were already treated with antihypertensive drugs. Sixty (67%) patients had decreased lower limb pulsations and femoral pulsations were absent in 19 (21%) patients. Other comorbid conditions included BAV in 39 (43%) and ventricular septal defect in 5 (6%) patients. Three patients (3%) had Turner's syndrome. Left ventricular hypertrophy was present in 49 (54%) patients. Ten patients (11%) underwent a previous cardiac surgery. In these patients, the CoA had initially insignificant severity in 6 and turned out to be overlooked in 4 cases.

The CoA was noted (paraductally) near the ductus arteriosus in most patients (64%), with 8% (preductally) in the aortic arch at the level of the left subclavian artery and 28% (postductally) distal in the proximal descending thoracic aorta. Poststenotic dilatation was noted in 19 (21%) patients and 7 (8%) patients had a hypoplastic isthmus. Aortic valve disease was found in 34 (38%) patients, 21 (23%) patients had aortic valve regurgitation, 8 (9%) had aortic valve stenosis and 5 (6%) combined aortic valve disease. Twenty-one patients (62%) out of those who had aortic valve disease had a BAV. Severe aortic valve regurgitation was present in 4 (4%) and severe aortic valve stenosis in 1 (1%) patients. All of these patients required a surgical replacement of the aortic valve. It is our policy to initially perform the coarctation repair to provide afterload reduction and separately carry out aortic valve surgery later. Four of these patients underwent replacement of their aortic valve after a median of 1.1 year (IQR 0.5–13.6 years) following their initial coarctation repair.

Most patients (57%) underwent reconstruction with an end-to-end anastomosis (Table 2). Left heart bypass was utilized in 9

**Table 3:** Postoperative outcome

Outcomes	Entire cohort (N = 90)
<b>30-Day outcomes</b>	
Mortality	1 (1)
Rethoracotomy for bleeding or tamponade	5 (6)
Vocal cord paralysis	2 (2)
CVA	0 (0)
Paraplegia	0 (0)
Renal failure	0 (0)
Respiratory failure	0 (0)
<b>Long-term outcomes</b>	
Long-term mortality	21 (23)
CVA	11 (12)
TIA	5 (6)
Myocardial infarction	3 (3)
Hypertension	69 (77)
Hypertension treatment	68 (76)
Any cardiac reintervention	31 (34)
Reinterventions for re-CoA	4 (4)
Other cardiac reinterventions	27 (30)

Values are presented as n/N (%).

CoA: coarctation of the aorta; CVA: cerebrovascular accident; TIA: transient ischaemic attack.

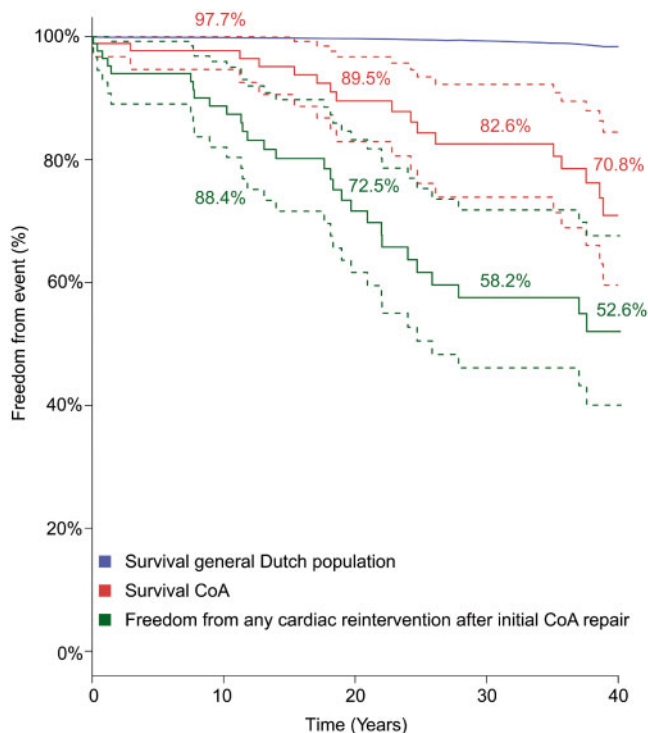
(11%) patients. There were no postoperative spinal neurological deficits and no cases of postoperative acute kidney insufficiency were reported. No patients experienced stroke or transient neurological deficit perioperatively. Two patients developed unilateral vocal fold paralysis due to lesion of the left recurrent laryngeal nerve with otolaryngology consultation and treatment.

In-hospital mortality was 1% (1 patient) (Table 3). This patient was a 39-year-old male with a known BAV and severely decreased left ventricular function due to previous anterior myocardial infarction. He was found to have a paraductal coarctation with a peak gradient of more than 80 mmHg on angiography. After open reconstruction with the end-to-end anastomosis he became haemodynamically unstable on the 16th postoperative day. He developed a hemothorax on the left side and subsequently died.

The median follow-up duration was 26.8 years (IQR 17.3–39.9 years). Twenty-one patients (23.3%) died during the follow-up period. The cumulative survival was 97.7%, 89.5%, 82.6%, 70.9% and 61.4% at 10, 20, 30, 40 and 50 years, respectively. Compared with age- and gender-matched Dutch population norms, the survival of patients with CoA undergoing open surgical repair was significantly impaired (Fig. 1).

Reintervention after CoA reconstruction was necessary in 4 (4%) of all in-hospital survivors. Two patients with initial end-to-end anastomosis and 2 patients with patch aortoplasty required a reintervention due to the recurrence of the coarctation. One of these patients with the initial patch aortoplasty was treated by surgical correction with interposition graft; the remaining 3 patients underwent stenting of the re-CoA. There were 15 (40%) cases of subsequent aortic valve replacement due to aortic valve disease (Fig. 2). Rates of freedom from any cardiac reoperation were 88.4%, 72.5%, 58.2% and 52.6% at 10, 20, 30 and 40 years, respectively. During the follow-up, 11 patients (12%) experienced a stroke and 5 patients (6%) had an incident of a transient ischaemic attack. Myocardial infarction occurred in 3 (3%) patients. The long-term follow-up showed that 69 (77%) patients still suffered





Number at risk after CoA repair:				
Mortality				
90	81	58	41	21
Reintervention				
90	67	41	26	12

**Figure 1:** Cumulative survival and freedom from any cardiac reintervention. Cumulative survival (represented by a red line) and cumulative freedom from reintervention (represented by a green line) after surgical repair of primary coarctation of 90 patients. The blue line is the survival of the normal population (matched for age and gender). Red dashed lines represent 95% confidence intervals of cumulative survival. Green dashed lines represent 95% confidence intervals of freedom from reintervention. CoA: coarctation of the aorta.

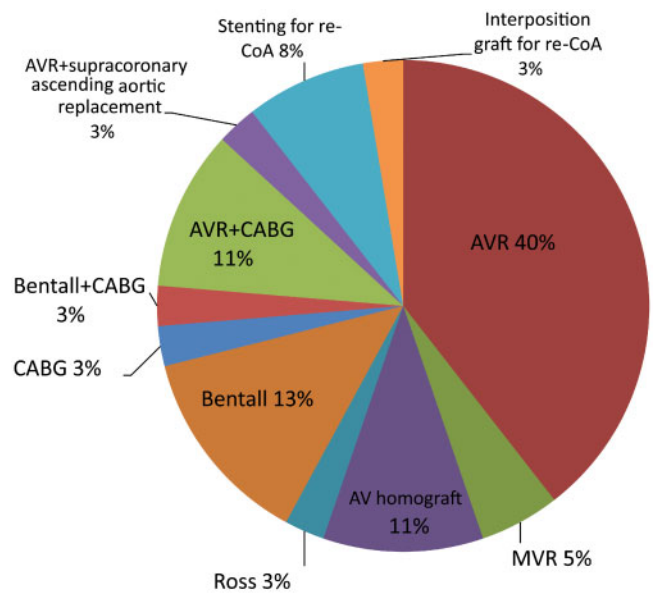
from hypertension and except for 1 patient they all complied with their antihypertensive treatment.

QoL could be assessed in 49 patients (82% of patients who received the questionnaire). All baseline characteristics were comparable between the respondents and those who did not return the questionnaire (Table 4). Compared with an age- and gender-matched Dutch population, male and female patients rated social functioning ( $P < 0.001$ ), mental health ( $P < 0.001$ ), vitality ( $P < 0.001$ ) and general health ( $P < 0.001$ ) significantly lower. Furthermore, they reported a higher bodily pain ( $P < 0.001$ ) (Table 5).

## DISCUSSION

The findings of the present study suggest that patients presenting later in life with CoA can successfully undergo complete resection, resulting in long-term durability. However, the perception that surgically repaired coarctation in young adults is a benign condition, should be met with caution as shown by our findings since the QoL of these patients is significantly impaired compared with the general population.

To the best of our knowledge, this is the largest adult cohort with very long-term outcomes of surgical correction of primary



**Figure 2:** Distribution of cardiac reinterventions (N = 38) after initial coarctation repair. AV: aortic valve; AVR: aortic valve replacement; CABG: coronary artery bypass grafting; CoA: coarctation of the aorta; MVR: mitral valve replacement.

CoA. Furthermore, our study spans over half century of operative history at the Erasmus University MC in Rotterdam. The median age of patients included in this study at the time of surgery was 24 years, with the youngest patient being 17 and the oldest 52 years old. Hence, these patients underwent their primary repair at a considerably older average age than the most reported data till now [10–13]. Consequently, the collateral vessels adjacent to the stenosis are more extensive and patients tend to present with other comorbidities that increase the challenges of surgical repair and risk of perioperative mortality and morbidity. Different authors described their experience with surgical management of CoA in adults. Yin *et al.* [14] reported their outcomes only up to 5 years in a group of 60 adolescent and adult patients and 28% of these patients underwent also a concomitant cardiac procedure. Hence, extra-anatomical bypass was the most common surgical technique while patch angioplasty was performed in 25% of all cases. They achieved satisfactory results with in-hospital mortality of 5.0% and paraplegia rate of 1.7%. In line with our findings, the 5-year survival was 98% in the isolated group, however, concomitant procedure was associated with markedly increased risk of mortality resulting in survival of only 82%. Furthermore, Yousif *et al.* [15] described their experience with treating 38 adult patients with interposition grafting of which nearly 45% were redo cases of previously repaired CoA. Their preferred approach included operative adjuncts such as distal aortic perfusion with left heart bypass and extracorporeal circulation with deep hypothermic circulatory arrest. They reported no in-hospital deaths or stroke. The long-term survival after 20 years was higher than in our cohort (94.7% vs 89.5%). Yet another report by Charlton-Ouw *et al.* [16] on 29 adult patients depicts their preference of interposition graft technique in case CoA is associated with thoracic aortic aneurysms. In addition, they describe their preference for distal aortic perfusion and deep hypothermic circulatory arrest, to protect organs from ischaemic consequences. There were no in-hospital deaths or strokes, and survival of 89% up to 17 years was similar to the results of the present study. Our findings mirror data from

**Table 4:** Demographic and clinical characteristics of SF-36 respondents

	Respondents in current study	Non-respondents in current study	P-value
Patients (n)	49	11	
Operation age (years), median (IQR)	22 (19–33)	25 (21–32)	0.32
Male gender, n (%)	29 (59)	6 (54)	>0.99
Asymptomatic, n (%)	24 (49)	5 (46)	>0.99
Hypertension, n (%)	49 (100)	11 (100)	
Hypertension treatment, n (%)	22 (45)	5 (46)	>0.99
Preoperative SBP (mmHg), median (IQR)	175 (151–188)	165 (150–170)	0.42
Preoperative DBP (mmHg), median (IQR)	91 (84–100)	95 (79–100)	0.49
COPD, n (%)	0 (0)	0 (0)	
Diabetes, n (%)	0 (0)	0 (0)	
Prior CVA, n (%)	1 (2)	0 (0)	>0.99
Prior cardiac surgery, n (%)	5 (10)	2 (18)	0.60
Associated anomalies, n (%)			
BAV	22 (45)	6 (55)	0.74
VSD	2 (4)	1 (9)	0.46
ASD	1 (2)	0 (0)	>0.99
PDA	1 (2)	0 (0)	>0.99
Hypoplastic isthmus	5 (10)	0 (0)	0.57
Hypoplastic aortic arch	0 (0)	0 (0)	
Poststenotic dilatation	15 (31)	0 (0)	0.051
Aortic regurgitation	15 (31)	3 (27)	>0.99
Aortic stenosis	10 (20)	1 (9)	0.67
Turner syndrome	1 (2)	0 (0)	
Location of coarctation, n (%)			
Preductal	3 (6)	0 (0)	
Juxtaductal	33 (67)	10 (91)	0.28
Postductal	13 (27)	1 (9)	

ASD: atrial septal defect; BAV: bicuspid aortic valve; COPD: chronic obstructive pulmonary disease; CVA: cerebrovascular accident; DBP: diastolic blood pressure; IQR: interquartile range; PDA: patent ductus arteriosus; SBP: systolic blood pressure; SF-36: Short Form 36-Item Health Survey; VSD: ventricle septal defect.

aforementioned studies describing the natural history of CoA after repair.

After repair of aortic coarctation, patients may develop restenosis, aneurysms and pseudo-aneurysms at the site of prior repair [17]. In our series no late aneurysm formation at the site of repair occurred. Therapy of either restenosis or (pseudo)aneurysm may consist of surgery, balloon angioplasty or stenting. The latest ACC/AHA 2018 guidelines for adults with congenital heart disease recommend either surgical repair or percutaneous catheter intervention for recurrent discrete CoA (Class I, Level of Evidence: B-NR) [6]. Furthermore, patients with coarctation are known for associated lesions, e.g. BAV and mitral valve abnormalities such as mitral valve stenosis which might require repair at some point. Hence, late problems such as reinterventions must be recognized. Brown *et al.* [12] reported freedom from any cardiac reoperation of 92.8% (10 years), 85.7% (20 years) and 76.6% (30 years). Compared with our results, they found lower reoperation rates at any time point. This can be explained by the high incidence of coexisting BAV in our cohort that contributes to the greater chance for reoperation in the future. In addition, aneurysm formation in the ascending aorta, proximal to the surgical site was remarkably prevalent. Available studies show wide reoperation rates with either surgical or catheter-based techniques ranging between 3% and 40% [10, 11, 18]. Our study reports 10.5% rate of reintervention due to restenosis. A repair technique other than simple or extended end-to-end anastomosis was independently associated with the need for reoperation of the aorta [10, 11, 18]. Subsequent aortic valve replacement was performed in 41.9% of

all reintervention cases. Our results reinforce the statement that end-to-end anastomosis is a durable operative technique and should be the technique of choice whenever possible.

Endovascular procedures have been used successfully for the treatment of CoA in predominately paediatric patients and little information exists regarding the endovascular repair in adult population. However, these procedures are rapidly becoming a popular way of treating many complicated conditions of thoracic aortic disease in the elderly. A recent multicentre study on endovascular treatment by Erben *et al.* [19] sought to examine short-term and intermediate results in an adult cohort with mainly redo patients (65.6%). Early mortality was 2% and postoperative complications included aortic dissections (3%), aortic ruptures (2%) and type IA endoleak (1%). Survival at 5 years was 89% and incidence of reintervention was 15% mainly due to residual claudication complaints and pseudoaneurysm formations. These limited data without extensive follow-up and no direct comparison to open repair suggest that the results need to be interpreted with caution.

Our results are consistent with the findings of others that late hypertension persists in patients with repaired CoA. Hager *et al.* [20] showed that even patients with optimal result after CoA repair were hypertensive at a rate of 55%. A systematic review by Canniffe *et al.* [21] has shown that the median prevalence of hypertension is 32.5% (range 25–68%) late after anatomically satisfactory CoA repair. As shown in this study, restenosis accounts only for a minority of cases of postoperative hypertension. These findings suggest that CoA might cause an irreversible effect on

**Table 5:** Quality of life comparison with matched Dutch general population

	Overall score (n = 49), mean ± SD	General Dutch population, mean ± SD <sup>a</sup>	Gender-matched comparison, P-value	General Dutch population, mean ± SD <sup>b</sup>	Age-matched comparison, P-value
PF	78.4 ± 26.1	83.3 ± 22.4	0.040	78.6 ± 11.1	0.007
SF	45.9 ± 13.8	84.3 ± 22.1	<0.001**	82.6 ± 3.6	<0.001**
RP	73.0 ± 39.5	76.7 ± 36.0	0.008*	72.0 ± 8.9	<0.001**
RE	76.2 ± 37.9	84.0 ± 32.4	0.007*	81.0 ± 3.2	0.007
MH	58.5 ± 10.3	77.0 ± 17.2	<0.001**	76.0 ± 1.7	<0.001**
VT	47.8 ± 11.0	68.7 ± 18.9	<0.001**	67.6 ± 2.8	<0.001**
BP	46.7 ± 11.5	75.0 ± 23.2	<0.001**	72.4 ± 4.01	<0.001**
GH	47.6 ± 11.2	70.9 ± 20.6	<0.001**	67.5 ± 6.4	<0.001**

\*Are no longer significant after Bonferroni correction, significant at P-value <0.05 level.

\*\*Significant after Bonferroni correction of 0.05/8 = 0.00625.

<sup>a</sup>Weighted average correcting for the proportion of males and females, respectively.

<sup>b</sup>Weighted average correcting for the proportion of patients in each age group, respectively.

BP: bodily pain; GH: general health; MH: mental health; PF: physical function; RE: role emotional; RP: role physical; SD: standard deviation; SF: social functioning; VT: vitality.

the upper body vasculature and early onset of hypertension might be related to changes in the vessel wall characteristics. Aortic stiffness develops and progresses with later correction of the CoA. These findings reflect a need for a more aggressive anti-hypertensive treatment, especially among the older patients who present with malignant hypertension at the time of surgery in order to decrease the higher chance of subsequent cardiovascular events.

Furthermore, this complex heart disease results not merely in the physical impairment but also in the emotional distress that may affect health-related QoL. As shown by this study different domains of QoL are significantly impaired when compared with the general population. The older average age at the time of surgery needs to be highlighted since these patients are more prone to numerous comorbidities and reinterventions. Bambul Heck *et al.* [22] confirm in their study that the increasing age is predictive of impaired QoL.

### Limitations and strengths

Although the current study is limited by its retrospective, mono-centric and observational nature, it is a consecutive series of patients. Furthermore, there was no comparative group of patients treated with endovascular techniques during the study period. These data cover the period where the patient care likely changed in ways not easily identified to confound the evaluated outcomes.

The strength of our results is that they provide accumulated data in a unique cohort of patients over 50 years of clinical experience. This study represents one of the largest populations of patients with primary adult CoA to be analysed and serves as a continuation of large contribution on long-term outcomes after surgical repair.

### CONCLUSIONS

Native adult aortic coarctation has low in-hospital morbidity and mortality when treated with an open surgical reconstruction; however, the risk of additional cardiac intervention and refractory hypertension remain important challenges during follow-up

of these patients. Furthermore, the health-related QoL is significantly impaired when compared with the general population. Life-long careful clinical follow-up and better adherence to secondary prevention are warranted for all patients after CoA repair. These findings may serve benchmarking with percutaneous treatment for adult CoA.

**Conflict of interest:** none declared.

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