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Surgical treatment of aortic coarctation in adults: Beneficial effect on arterial hypertension

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Abstract

Background: *The aim of this study was to determine the outcome after surgical repair of aortic coarctation in adults, analysing its effect on arterial blood pressure.*

Methods: *Twenty-five adults (9 women, 16 men), mean age 43.4 years (19 to 70 years), underwent aortic coarctation surgical repair. All patients suffered from preoperative hypertension. Mean blood pressure was 182/97 mm Hg. Sixteen (64%) patients demonstrated reduced load capacity. Operative technique was resection and end-to-end anastomosis for 5 patients (20%), interposition of a Dacron-tube graft for 3 patients (12%), Dacron-patch dilatation was performed in 7 (28%) patients, and in 10 (40%) patients we performed an extra-anatomical bypass graft.*

Results: *Early mortality occurred in 1 patient (4%). The mean blood pressure was reduced [systolic 182 mm Hg* vs. *139 mm Hg ($p < 0.001$), diastolic 97 mm Hg* vs. *83 mm Hg ($p < 0.001$)] in all patients. In 12 patients, blood pressure normalized immediately after surgery, in 7 patients it remained slightly elevated (systolic blood pressure between 140–160 mm Hg), and 1 patient suffered from prolonged arterial hypertension. Preoperatively, all patients were treated with antihypertensive drugs. Eleven of 20 patients received long-term medication during follow-up. In the remaining 4 patients, medication lists were unobtainable in retrospect. The mean follow-up was 7.1 years (min. 1.0 years; max. 16.6 years). One patient (5%) died from cardiac failure 12.4 years after the operation. On average, the New York Heart Association (NYHA) class was improved by 0.92.*

Conclusions: *The surgical repair of aortic coarctation in adults can be performed with low surgical risk. Surgery reduces hypertension and permits more effective medical treatment.* (Cardiol J 2008; 15: 537–542)

Key words: aortic coarctation, coarctation of the aorta, arterial hypertension, persisting hypertension

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Introduction

Aortic coarctation (CoA) is a congenital anomaly defined as a stenosis of the aorta in the area of the ligamentum arteriosum. In up to 30% of all cases, it coincides with other malformations, e.g. bicuspid aortic valve or subvalvular aortic stenosis. According to the age of the patient at first presentation, CoA is commonly divided into infantile or adult form. Whereas the infantile form can lead to life-threatening situations in neonates after physio-

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logical occlusion of the ductus arteriosus, the adult form can remain asymptomatic. Therefore, CoA displays various symptomatic features according to the patient's age and triggers a range of surgical problems. Characteristic clinical symptoms accompanying the adult form of CoA are hypertension of the upper part of the body, headache, frequent epistaxis as well as paresthesia and/or claudication of the lower extremity [1].

Uncorrected, CoA has an unfavourable prognosis. Long-term progression corresponds in its untreated form with atherosclerosis, myocardial infarction due to accelerated angiosclerosis and cardiac hypertrophy, cardiac insufficiency, aortic rupture with or without dissection, bacterial endocarditis and cerebral hemorrhage. Mean life expectancy is described to be between 35 and 45 years [1–7]. To avoid hypertension-related complications, operative intervention is the treatment of choice. Surgical reconstruction of CoA has been successfully performed since 1944 [8]. It is applied to every age. However, surgery of adults comprises — besides general operative and anesthetic risks — further specific disadvantages, i.e. calcifications within the coarctation area, aneurysms and large intercostal arteries. The aim is to obtain permanent normotension or a notable reduction of the blood pressure gradient. Besides an end-to-end anastomosis, further operative techniques, such as Dacron-patch plasty, have been developed. With complex CoA, extra-anatomic bypass grafts between the ascending or left subclavian and the descending aorta are effective techniques [9–11]. Especially in adult patients with CoA repair, persisting arterial hypertension may occur despite satisfying surgical results [6, 11, 12].

Methods

Between August 1985 and August 2006, 25 patients (9 female, 16 male) underwent surgical repair of CoA as adults. Their mean age was 43.4 years (range: 19 to 70 years), mean age at the first diagnosis was 39 years (min. 15 years, max. 65 years). The mean interval between diagnosis and surgical intervention was 1.6 years (min. 0 years, max. 13 years).

In 11 of 21 patients (52.4%), CoA was diagnosed as an incidental finding (in 4 patients, retrospective diagnostic review was impossible). All patients showed hypertension of the upper extremities. Preoperative blood pressure of the upper extremities was 182/97 mm Hg, on average (systolic between 140 and 230 mm Hg, diastolic between 70 and 110 mm Hg). The maximum pressure gradient ranged between 35 and 110 mm Hg (mean 62.7 mm Hg). Two patients (8%) presented with recurrent CoA after a previous repair: one patient had obtained a Dacron-patch plasty 13 years ago at the age of 26, and the second patient was at first treated with a percutaneous, transluminal angioplasty 9 months before the operation. A preoperatively reduced load capacity was diagnosed in 16 patients (64%): 4 patients were classified as New York Heart Association (NYHA)-class I, 9 as NYHA-class II, 2 as NYHA-class II–III and 1 patient as NYHA-class III. Eight patients were asymptomatic, and 1 patient could not be classified in retrospect. Ten patients (40%) showed minor valve disease not necessitating surgical intervention. Cardiac arrhythmias such as supraventricular and ventricular were detected in 7 patients (28%). Rib erosions were visible on the chest roentgenograms of 19 patients (76%). As a surgical technique, end-to-end anastomosis was performed in 5 patients (20%), Dacron-patch plasty in 7 (28%) early in the series, resection and interposition of a Dacron-tube graft in 3 patients (12%). An extra-anatomical bypass graft was placed in 10 patients (40%) with 5 patients receiving an ascending-descending aorta and 5 patients receiving a subclavian artery-descending aorta bypass graft. The operating time ranged between 105 and 295 min (mean: 194 min).

Results

Early mortality occurred in 1 patient (4%) as a result of suture dehiscence. This patient was immediately re-operated, but died from the sequele of hemorrhagic shock. Another patient developed bleeding in the chest wall on the eleventh postoperative day. After surgical revision the further postoperative course was uneventful.

Postoperative mean systolic and diastolic blood pressures were reduced (systolic 182 *vs.* 139 mm Hg ($p < 0.001$), diastolic 97 *vs*. 83 mm Hg ($p < 0.001$) in all patients. Of the 25 patients with preoperative hypertension, 12 were normotensive after surgery, in 7 patients blood pressure remained slightly elevated (systolic blood pressure between 140 and 160 mm Hg), and 1 patient had prolonged hypertension (Table 1). For the remaining patients, valid data were not available. Because of a tendency for tachycardia shortly after the operation in association with still elevated blood pressure, beta-blockers were given as a first choice in all patients. Preoperatively, all patients were treated with antihypertensives. Postoperatively, 11 of 20 patients received long-term medication during follow-up (Table 1). The

Table 1. Aortic coarctation repair: pre- and postoperative blood pressure, pressure gradient and medication.

Number	Systolic/diastolic blood pressure [mm Hg]: Upper extremity		Pressure gradient [mm Hg] between upper and lower extremities		Medication	
	Preoperative	Follow-up	Preoperative	Follow-up	Preoperative	Follow-up
1	160/110	120/75	35	9	B; D	B
2	165/85	130/85	40	10	B; C	
3	230/100	135/80	110	5	B; C; D	B
4	205/110	185/100	95	20	B; C; D	B; C
5	140/100	–	45	–	B; D	–
6	180/95	125/75	55	0	B; C; D	
7	180/90	–	35	–	B; C, D	–
8	170/95	130/80	50	12	B; C	B
9	190/95	–	85	–	B	–
10	185/95	–	35	–	B; D	–
11	185/100	–	80	–	B; C	–
12	170/95	165/85	65	0	A; B; D	B; D
13	180/100	130/85	55	5	A; C, D	
14	195/105	170/95	80	15	A; B; D	A; B
15	180/95	125/70	70	5	B; C	
16	195/100	135/80	75	7	A; C, B	
17	175/90	130/80	60	5	B; C; D	B
18	200/100	170/90	90	10	A; B; D	A; B
19	190/90	160/90	55	10	B; C; D	B; C
20	195/100	135/85	75	5	B; C; D	B
21	170/105	125/80	40	0	B; C; D	
22	165/70	125/85	40	0	B; C	B
23	170/90	125/80	45	0	A; B; D	
24	200/110	135/85	100	5	A; B; C; D	
25	180/95	130/80	65	5	A; B; D	

A — angiotensin-converting enzyme inhibitors; B — beta-blockers; C — calcium antagonists; D — diuretics; – no follow-up data

mean postoperative follow-up period was 7.1 years (min. 1.0 years; max. 16.6 years). One patient (5%) died 12.4 years after the operation from cardiac failure, late mortality therefore being 5%.

Nine patients (45%) improved their NYHA--classification (5 patients by 2 classes; 4 patients by 1 class). In 9 patients (45%) load capacity remained unchanged. Two patients (10%) deteriorated at time of follow-up by 2 classes (first patient: age at operation: 48 years, follow-up 11.5 years, suspicion of dilated cardiomyopathy — second patient: age at operation: 61 years, follow-up 12 years, developed a valvular aortic stenosis of a bicuspid aortic valve). On average, the NYHA classification was improved by 0.92 NYHA-classes. No data were available for the remaining patients.

At the time of follow-up only 5 of 20 patients (40%) displayed prolonged hypertension with indication for a reduced combined antihypertensive therapy. In 6 of the remaining 15 patients, normotension was achieved by administration of beta--blockers. Nine patients received no further antihypertensive medication (Table 1).

Discussion

Aortic coarctation causes a pressure gradient between the upper and lower extremities. With its characteristic symptoms, CoA is often diagnosed during infancy and treated surgically accordingly. The combination of hypertension of the upper part of the body with weakened pulses of the groin or foot is typical. However, a hemodynamically relevant CoA is often diagnosed only in adults [11, 13–15]. Physical development proceeds perfectly normally in most cases. Arterial hypertension is repeatedly diagnosed only as an incidental finding, as in 11 of our 21 patients (52.4%). Those patients were treated with suspicion of essential hypertension until a congenital CoA was finally diagnosed.

Surgical indication is defined by the diagnosis of CoA alone. The adult form of CoA is accompanied by a limited life expectancy in its natural course. Most patients die before their fortieth birthday. Cardiac defects, aortic rupture, bacterial endocarditis or endarteritis with their complications as well as cerebral hemorrhage are the most common causes of death in connection with CoA [2, 5, 15].

Surgical correction is usually performed via a left-sided posterolateral thoracotomy. The most common operative technique during infancy comprises a resection of the stenosed segment with an end-to-end anastomosis during aortic clamping. This operative treatment was established by Clarence Crafoord in 1944 [8]. During clamping, the blood supply of the lower part of the body is maintained by collateral circulation only. The most dreaded complication in CoA surgery is postoperative paraplegia. The frequency of occurrence correlates with the duration of clamping and the resulting blood pressure in the lower half of the body, as well as with age [16]. It can be alleviated by distal perfusion strategies, especially in the adult. Operative mortality in adult patients is relatively low, being about 4.5% [4, 8, 11, 17].

In older patients, resection of CoA or re-operation of recurrent CoA is technically more difficult and often even impracticable due to the poor quality of the vessel walls, rigid vessel walls with calcification in the CoA area, and aneurysmatic alterations of intercostal arteries [13, 17–19]. Because of these problems, interposition of a prosthesis is usually feasible in the adult. Dacron-patch plasty has become obsolete because of dismal late results [20, 21]. Extra-anatomic bypass graft techniques with the optional use of extracorporeal circulation are interesting alternatives [13, 21, 22].

During indirect isthmus-plastic surgery according to Vossschulte, a Dacron-patch is used to augment the stenotic aortic segment. This technique requires only a short-distance dissection of the aorta, without impairment of the collateral vessels, theoretically minimising the risk of paraplegia [23, 24]. Despite the frequently described risk of postoperative formation of aneurysms as sequele of the above-mentioned technique, it is nevertheless still applied in certain cases where long-distance dissection is to be avoided [21, 25]. By using autogenous arterial vessels, some authors emphasize the reduced risk of re-stenosis or development of aneurysms compared to the application of Dacron as patch material [25].

For re-operations or in complex CoA, extra-anatomical bypass grafts can be employed [2, 9, 11, 13, 14].

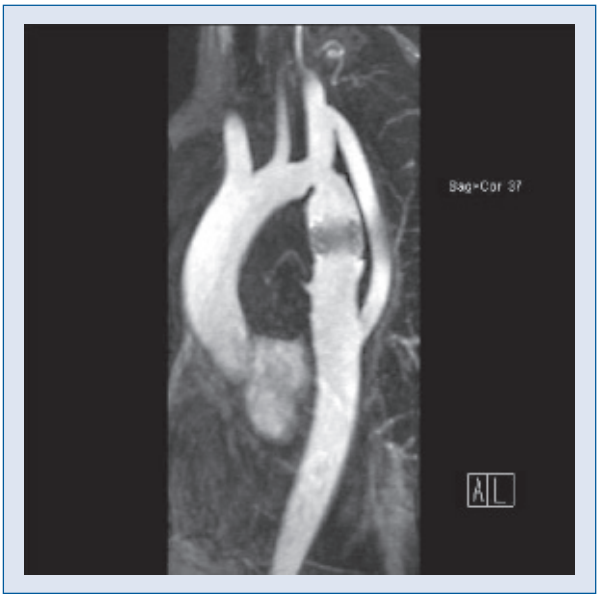

Figure 1. Left subclavia — descending aortabypass.

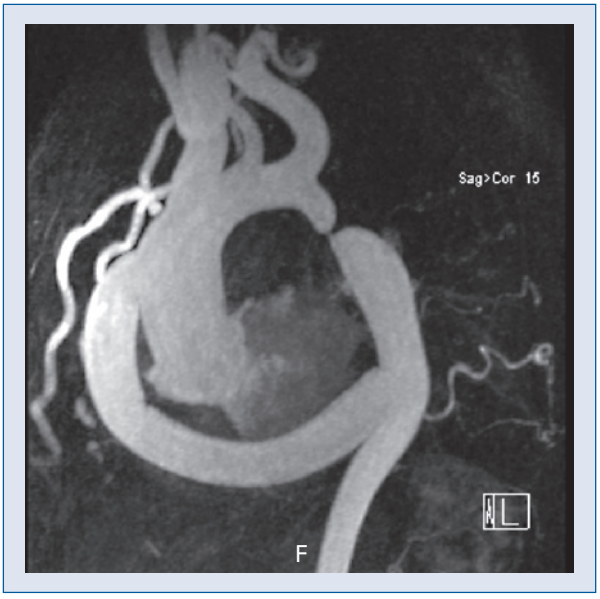

Figure 2. Ascending aorta — descending aortabypass.

A subclavian artery-descending aorta-bypass is administered via left-sided thoracotomy. The subclavian artery has to comprise at least 60% of the lumen of the descending aorta at the level of the diaphragm. An ascending aorta-descending aorta bypass is usually performed via median sternotomy (Fig. 1, 2). Depending upon the access to the descending aorta, extracorporeal circulation may be necessary.

One of the most serious complications after surgical repair of CoA is the incidence of re-stenosis.

The risk of re-coarctation after surgical treatment in adults is described between 0% and 9% [13, 20]. As an indication for re-intervention we defined a maximum gradient > 20 mm Hg between the right arm and leg or as measured during cardiac catheterization. Doppler sonography as a single means to determine the remaining gradient or the development of a recurrent gradient after CoA repair proved to be unsuitable [26]. Due to the advancing progress of interventional cardiology, re-stenoses or remaining stenoses can be dilated with or without deployment of a stent graft. This has become an attractive alternative to surgical interventions [27–30]. Long-term results remain to be seen.

Despite positive postoperative results (no gradient), a prolonged elevation of blood pressure, now in the lower as well in the upper body half, is found in about 15–25% of operatively treated patients, especially in those treated later in life. To this day, operation at a young age is the most important prophylaxis against future hypertension [4, 28, 30]. In 5 of our patients, hypertension persisted postoperatively.

As all patients responded with tachycardia shortly after the operation in association with still elevated blood pressures, they were treated with beta-blockers routinely. Pathophysiologically this phenomenon may be explained by still severely diminished peripheral vascular resistance because of long-standing inflow stenosis of the vascular bed.

Preoperatively all 20 patients had been treated for arterial hypertension of unknown causes for various periods of time. After a mean follow-up of 7.1 years, only 5 of 20 patients (25%) showed a persisting hypertension, which could not be reduced to normotension despite medication. The remaining 15 patients showed a normotensive blood pressure. Nine patients (45%) were not treated with antihypertensive drugs. Medication could be distinctly reduced in all patients (Table 1). Our patient population exhibited a postoperative improvement of the functional condition according to the NYHA-classification and can be compared to the data published by Cohen et al. [4].

Therefore, a competent lifelong medical surveillance of patients with CoA repair is mandatory. To gain a detailed record of postoperative alterations, such as re-stenosis, development of aneurysms or the progression of valve defects, follow-up examinations at regular intervals are necessary. For imaging, high-resolution techniques, e.g. echocardiography computed tomography (spiral-CT) and magnetic resonance imaging (MRI), are suitable means (Fig. 1, 2) [24, 31–33].

This supervision should be based on close cooperation between specialised cardiologists with expert experience in the area of congenital cardiac defects as well as cardiac surgeons. Some countries have established particular task forces [34].

Aortic coarctation often occurs in combination with aortic valve defects, mainly a bicuspid aortic valve. However, few patients need aortic valve replacement before adulthood. In the long-term, however, aortic valve disease remains a significant cause for morbidity and mortality in patients with CoA [35]. Various studies describe the improvement of life expectancy after corrective surgery. Nevertheless, normal life expectancy is rarely achieved [4, 29, 35, 36]. In this study, coexistent aortic valve defects were diagnosed in 8 patients (40%). Not all of the valves had to be treated surgically at the time of CoA repair. During the follow-up period, however, 5 patients (20%) underwent an aortic valve replacement, a higher rate than that described by other authors [4, 35]. Associated valve disease is one of the major reasons for recommendation of a strict endocarditis prophylaxis [11, 29].

According to various studies, perioperative mortality for adult CoA repair is between 0% and 4.5%. The perioperative mortality in this study was 4%. Late mortality after a median follow-up time of 7.1 years was 5% (1/20) and therefore lower than in the above-mentioned studies.

Limitation of the study

The limitations of this study are its retrospective nature, the small number of patients, and the lack of objective exercise-induced hypertension testing.

Conclusions

Overall, the present study shows that surgical repair of CoA in adults reduces arterial systolic and diastolic hypertension and increases load capacity. These operations can be carried out with a low early and late morbidity and mortality rate. The CoA repair simplifies the adjustment of blood pressure and is often associated with a reduction of antihypertensive agents. After surgical therapy, a subsequent lifelong medical follow-up of the patients in specialised centres is mandatory.

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