

SURGERY FOR COARCTATION OF THE AORTA: LONG-TERM POST-OPERATIVE RESULTS

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A b s t r a c t

In this retrospective study, the results of surgery for coarctation of the aorta in 376 patients treated at the Centre of Cardiovascular Surgery and Transplantation in the 1978/2001 period were evaluated. The average age at surgery was 7.2 ± 8.9 years and 41 patient were younger than 3 months. The methods most frequently used included resection with end-to-end anastomosis (189 patients), patch-graft aortoplasty (136) and subclavian flap aortoplasty (24). The long-term follow-up involved 314 patients; in 248, the outcomes of treatment were good. These patients were free of recoarctation and showed normal blood pressure values. The patients who had surgery at less than 1 month of age and those in whom a residual arm/leg gradient remained after the primary operation were at risk of disease recurrence. A total of 24 (7.6%) patients underwent reoperation for recoarctation; there was no relation to the initial surgical method. Persistent hypertension was recorded in 15 (4.8%) patients although they had good anatomical repair of the defect and were treated with anti-hypertensive medication. The development of aneurysm at the site of patch aortoplasty performed according to Vosschulte was the cause of reoperation in five patients; this method was later abandoned. The results showed that the best outcomes are achieved in children treated at pre-school age, which is associated with the lowest risk of late complications. It was concluded that, because of potential development of recoarctation, aortic aneurysm or late hypertension, all patients with coarctation repair should be followed up for the rest of their lives.

Key words

Coarctation of the aorta, Recoarctation, Hypertension, Aortic aneurysm

INTRODUCTION

Coarctation of the aorta is the fifth most common congenital heart defect, accounting for 5 to 9%, with male infants being affected twice or three-times more often than female babies. It is characterised by a significant narrowing of the proximal section of the descending thoracic aorta. Isolated juxtaductal coarctation, usually asymptomatic in childhood, is the cause of secondary hypertension, which is a serious complication that limits patient's life expectancy. The average survival in non-treated patients is about 35 years, with only 20% reaching the age of 50 years (1, 2, 3). The most frequent causes of death include heart failure (18%), subarachnoid haemorrhage (11%), aortic rupture or dissection (23%) and bacterial endocarditis (22%) (2, 4). Infancy coarctation is usually

preductal, with a hypoplastic aortic arch and a wide arterial duct, and is often (in 35 to 60%) associated with other heart defects (5, 6). If this coarctation is very narrow or concomitant cardiovascular disorders do exist, ductal occlusion, if it occurs, may result in congestive heart failure. If this critical condition cannot be controlled medically, it should be treated surgically as early as possible.

The objective of surgical repair is to achieve, by resection or aortoplasty, normal perfusion in the part of the body distal to the aortic stenosis, and thus provide conditions for blood pressure normalisation and cardiac compensation. The most commonly used methods include resection with end-to-end anastomosis, patch-graft aortoplasty, subclavian flap aortoplasty, direct aortoplasty by transverse suture repair, extended resection with end-to-end anastomosis and resection with end-to-end conduit interposition (4, 7, 8, 9).

The major complications of the long-term, post-operative course arise due to residual or recurring coarctation, persistent hypertension or development of aortic aneurysm (1–10). The aim of this retrospective study was to review the results of surgical treatments carried out in our Centre in the period from 1978 to 2001 and to evaluate them in relation to the age of patients, type of coarctation, year of surgery and the method used.

MATERIALS AND METHODS

The clinical records of 376 patients surgically treated for coarctation of the aorta at our Centre between January 1978 and December 2001 were reviewed. The average age at the time of surgery was 7.2 ± 8.9 years (range, 2 days to 44 years). There were 41 infants younger than 3 months (10.9%), 301 children younger than 15 years (80.1%) and 34 adult patients (9.0%). The male-to-female ratio was 1.81:1. The surgical procedures used (*Fig. 1*) included resection with end-to-end anastomosis in 189 (50.2%) patients, patch aortoplasty in 136 (36.1%), subclavian flap aortoplasty in 24 (6.5%), direct aortoplasty in 18 (4.8%), resection with conduit interposition in eight (2.1%) and prosthetic bypass in one (0.26%) patient. The long-term results were evaluated in 314 patients; 62 were excluded because they were lost to follow-up or this was too short.

The follow-up periods ranged from 1 to 24 years (mean \pm SD, 8.1 ± 6.1 years). The first examination was at 6 months after surgery and then approximately every two years. It included blood pressure measurement on the upper and lower extremities, X-ray and echocardiography. When aortic aneurysm was suspected or the findings were not consistent, examination by computed-tomography or nuclear resonance imaging of the thorax was carried out. The presence of residual or recurrent coarctation was suspected in the patients in whom arm blood pressure was higher by 20 mm Hg or more than leg blood pressure. Indications for reoperation for recoarctation included arm/leg pressure gradients higher than 30 mm Hg and hypertension. In adults, hypertension was shown by blood pressure higher than 140/90 mm Hg and, in children, it was blood pressure higher by 20% than normal pressure for their age category.

Values within each category were compared and the differences were statistically analysed using Fischer's exact test. A *P* value < 0.05 was considered statistically significant.

RESULTS

Out of the total 376 patients reviewed, 11 (2.92%) died in the early post-operative period. Ten of them were younger than 3 months and had critical

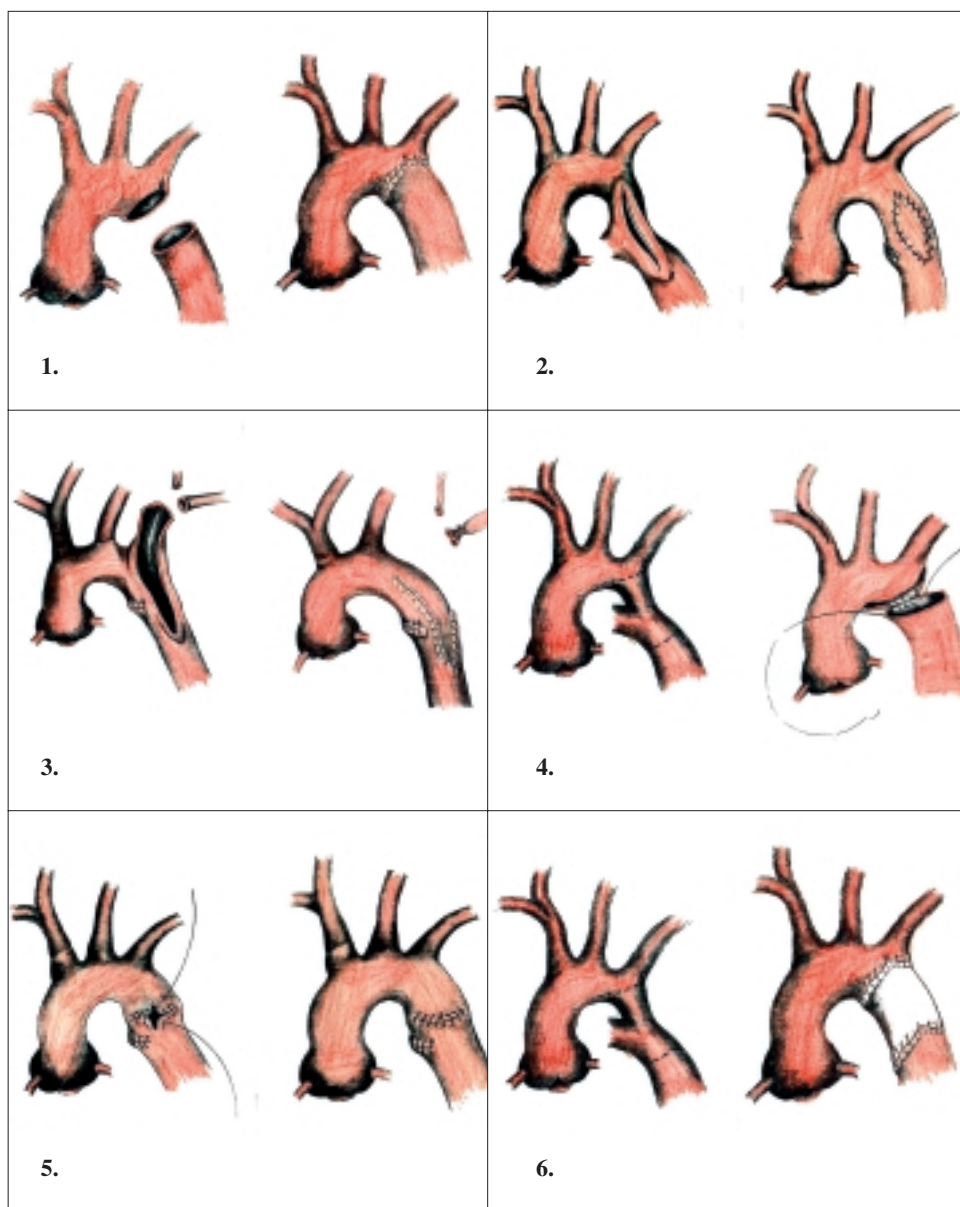


Fig. 1.

Schematic presentation of the surgical procedures used. 1. Resection with end-to-end anastomosis; 2. Patch-graft aortoplasty; 3. Subclavian flap aortoplasty; 4. Extended resection with end-to-end anastomosis; 5. Direct aortoplasty by transverse suture repair; 6. Resection with end-to-end conduit interposition.

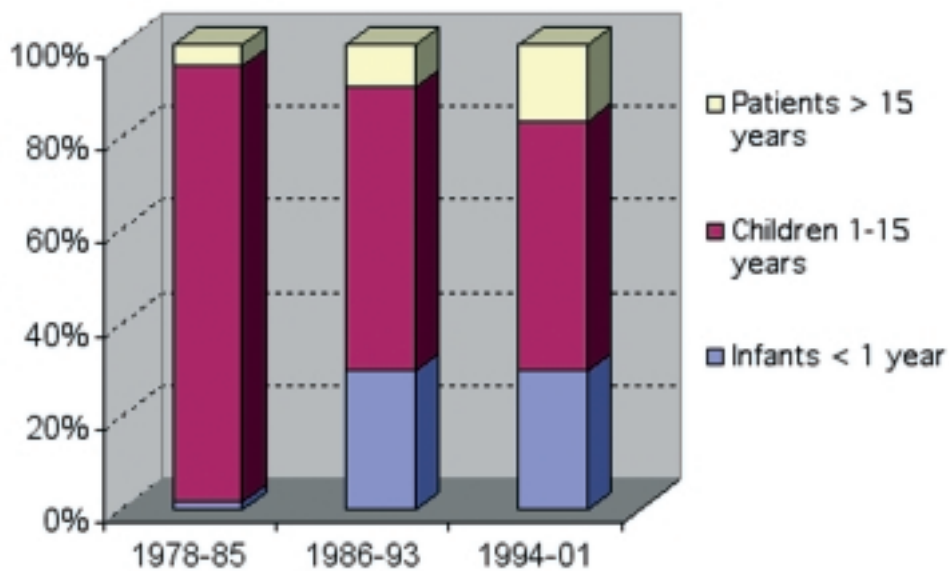


Fig. 2.
Percent of patients treated, as assessed by age categories

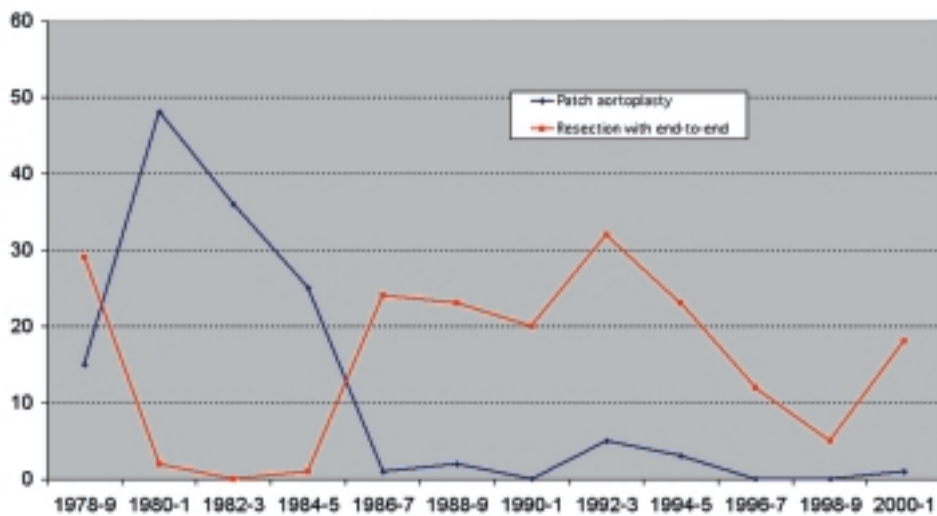


Fig. 3.
Use of the two major surgical procedures in each year of the follow up.

coarctation due to severe isthmus and arch hypoplasia or coarctation associated with other cardiovascular diseases, namely, ventricular septal defect (two patients), double outlet right ventricle (one), common arterial trunk (one) and severe isthmus and arch hypoplasia (three patients). One child died during the radical correction of an associated congenital defect at the age of 20 months, two days after surgery for coarctation of the aorta. The age distribution of the patients treated in the period from 1978 to 2001 and use of two major surgical procedures are shown in *Figs 2 and 3*, respectively.

In the group of 314 patients at long-term follow-up, three died following further surgery for associated intracardiac pathology at 4, 11 and 12 years, respectively, after the initial operation for coarctation.

A total of 248 (79%) from the 314 followed-up patients had normal blood pressure values and no or low arm/leg gradients; this was regarded as a very good outcome. Fifty one (16.2%) patients had the arm/leg gradient higher than 20 mm Hg. Out of these, 27 (8.6%) were followed up for recoarctation that was not clinically significant to an extent requiring reoperation, and 24 patients with hypertension and a gradient higher than 30 mm Hg underwent repeat surgery. The average interval between operations was 7.1 ± 5.3 years. None of these 24 (7.6%) patients died; two later developed signs of recoarctation and 22 showed good outcomes of the repeat surgery. More data on the patients undergoing reoperation for recoarctation are given in *Table 1*.

Persistent hypertension after good anatomical repair of coarctation was recorded in 15 (4.8%) patients (*Table 2*). Five patients, initially undergoing patch aortoplasty, had repeat surgery for aortic aneurysm, with two being emergency procedures because of signs of aorto-bronchial fistulas. All the surgically treated patients survived with a record of satisfactory health status. Three patients diagnosed with aneurysm were indicated for stent-graft insertion, which was carried out in the Centre for Vascular Intervention, Regional Hospital, Ostrava. Four patients with the signs of developing aneurysms remained under clinical observation.

DISCUSSION

RECOARCTATION

Patients operated on before one year of age are at higher risk of recoarctation than the older patients. The incidence reported is ranging from 8 to 54% and had been higher in the earlier period (4, 7). The development of recoarctation is attributed to the insufficient growth of anastomosis, presence of ductal tissue that, in critical coarctation, tends to retract, or technical faults such as insufficient resection or a wrong estimate of patch size (10,11). It is probable that many recoarctations are in fact residual coarctations, particularly in patients operated on at the time when fine, absorbable suture materials were not available (12). This is

Table 1

Percent of reoperations for recoarctation in patients categorised according to the period, age and surgical technique used at the primary surgery

Reoperation for recoarctation		
Period		
1978–1985	11.0%	(16/146)
1986–1993	5.1%	(6/118)
1993 – 2001	4.0%	(2/ 50)
Age category		
Newborns, less than 1 month	36.4%	(4/ 11)
Children, 3 months to 15 years	6.8%	(19/279)
Adults, more than 15 years	4.1%	(1/ 24)
$P < 0.001$		
Surgical procedure		
Resection with end-to-end anastomosis	4.2%	(8/189)
Patch-graft aortoplasty	7.3%	(10/136)
Subclavian flap aortoplasty	8.3%	(2/24)
Direct aortoplasty	5.6%	(1/ 18)
$P = 0.381$		

Table 2

Proportions of patients with persistent hypertension in relation to the age at primary surgery

Age at surgery	Persistent hypertension	
Younger than 6 years	2.1%	(4/189)
Six to 15 years	5.0%	(5/101)
Older than 15 years	25%	(6/ 24)
$P < 0.001$		

in agreement with our observations. In the patients who underwent reoperation for recoarctation, as compared with the patients not requiring repeat surgery, there was a significantly higher proportion of those who showed substantial residual stenosis after the primary surgery (29.8% against 3.1%). Two thirds of our patients with repeat surgery underwent the primary surgery between 1978 and 1985. The patient's age at primary surgery was also important; we found the highest proportion of reoperations for recoarctation in the youngest age category (under 1 month). However, the method used for primary surgery did not play any significant role.

PERSISTENT HYPERTENSION

Hypertension is a frequent complication of coarctation of the aorta. Its persistence is related to the patient's age at the primary operation, older age being associated with a higher probability of persistent hypertension even in patients free of recoarctation. This is explained by the abnormal function of baroreceptors and an increase in peripheral vascular resistance in the upper extremities that developed before the coarctation has been corrected surgically and may persist for years afterwards (13). In patients who have undergone surgery for coarctation in adulthood and are free from recoarctation, persistent hypertension is reported to be present in 25% to 53% (4, 14). Our findings agree with this data because we found persistent hypertension in 25% of the patients operated on after the age of 15, and hypertension on discharge from hospital was a risk factor for late hypertension. All hypertensive patients require anti-hypertensive medication but, even if on this therapy, they are still at an increased risk of coronary heart disease, cerebral haemorrhage, aortic aneurysm or dissection development.

SURGICAL PROCEDURES

Ideally, the surgical procedure should remove all "ductal" tissue and provide a broad anastomosis that retains its growth potential. In patients with isolated coarctation of the aorta, a simple resection of coarctation combined with the use of absorbable suture materials achieves good outcomes. Recoarctation has been reported to occur in 3 to 8% of these patients; in our group it was 3.8%.

In newborns and infants with hypoplastic aortic isthmus, simple resection does not treat the aortic stenosis completely, and this remains a risk factor for the development of recoarctation in 10% to 54% of these patients. Subclavian patch aortoplasty has not provided better results; coarctation has been reported to recur in 8% to 45% (8, 15, 16). In addition, deficient growth of the left upper extremity with decreased muscle force has been described. The complete removal of pathological ductal tissue and hypoplastic isthmus has been achieved by extended

resection with end-to-end anastomosis. This procedure was used in 11 of our patients and they all remained free from recoarctation.

Direct aortoplasty was indicated in 18 children in whom the anatomy of this defect allowed for this intervention. One patient underwent reoperation for recoarctation and 17 showed good outcomes. The results of direct aortoplasty are seldom reported in the literature.

After coarctation management with plastic patch aortoplasty according to Vosschulte, a true aneurysm or pseudoaneurysm has been reported to develop at the surgery site in 5% to 38% of the patients. The true aneurysm is usually localised opposite to the patch and the causes of its development include a patch more rigid than the vessel wall, presence of persistent hypertension or excessive resection of the aortic rim (4,14,17,18,19). The patients affected are at risk of sudden death due to rupture of the aneurysm or creation of an aorto-bronchial fistula. The latter condition was the reason for emergency surgery in two of our patients. After the year 1986, this method was used only occasionally in our group.

In older children with a long aortic stenosis, sufficient mobilisation of the aorta is not possible and the segment resected has to be replaced with a prosthesis. This approach was used in eight of our patients. In two patients, after they ceased to grow, the prosthesis had to be replaced by another with a larger diameter.

It can be concluded that surgical repair of coarctation of the aorta has a good outcome in almost 80% of the patients treated. Both mortality and late morbidity are affected by the patient's age at the time of surgery. In newborns and infants, who are at high risk of recoarctation, surgery is indicated only if cardiac failure cannot be controlled by medication. Pre-school children are at the lowest risk of recoarctation and persistent hypertension and, therefore, the age recommended for the surgical treatment of coarctation is between three and six years.

None of the surgical techniques available for management of coarctation has general applicability. Even though resection can be used in most patients, each case should be evaluated individually in order to choose the optimal method. Each patient requires a long-term follow-up because post-operative complications are frequent and their number may increase with years after surgery.

KOARKTACE AORTY: CHIRURGICKÁ LÉČBA A DLOUHODOBÉ POOPERAČNÍ VÝSLEDKY

S o u h r n

Autoři v retrospektivní studii uvádějí výsledky operací koarktace aorty u 376 nemocných, kteří byli operováni na kardiochirurgickém pracovišti v Brně v letech 1978–2001. Průměrný věk v době operace byl 7,2(8,9 roku, 41 operovaných bylo mladších 3 měsíců. Nejčastěji použitými operačními postupy byla resekce koarktace aorty (189), plastika koarktace záplatou dle Vosschulte (136) a Waldhausenova plastika (24). Dlouhodobé výsledky jsou hodnoceny u 314 operovaných. Dobrého výsledku operace bylo dosaženo u 248 (79%) nemocných, kteří jsou bez rekoarktace a mají normální hodnoty krevního tlaku. Reoperováno pro rekoarktaci bylo 24 nemocných (7,6%), rizikovým faktorem ve vztahu k rekoarktaci byl věk do 1 měsíce a reziduální gradient mezi horními a dolními končetinami po první operaci ($P(0,001)$). Chirurgická metoda neměla u našeho souboru statisticky významný vliv na incidenci rekoarktace. Hypertenze při medikaci a dobré anatomické úpravě vady přetrvává u 15 operovaných (4,8%) a její výskyt narůstal se zvyšujícím se věkem nemocných v době operace ($P(0,001)$). Rozvoj výdutě aorty v místě rozšiřující plastiky záplatou dle Vosschulte byl příčinou reoperace u 5 nemocných a tuto metodu dále neužíváme. Z výsledků vyplývá, že nejvhodnější dobou pro operaci koarktace aorty je období předškolního věku, kdy je riziko pozdních komplikací nejnižší. Vzhledem k možnosti vzniku rekoarktace, výdutě aorty a pozdní hypertenze však doporučujeme trvalou kardiologickou a kardiochirurgickou dispenzarizaci všech operovaných.

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