CLINICAL STUDY

Improved patient survival following surgery for coarctation of the aorta

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Abstract

Background: **We conducted a retrospective review of children undergoing surgery for coarctation of the aorta in our institution over the last ten years with the aim of evaluating overall patient survival as well as detecting factors affecting it. We tried to identify the risk factors for mortality.**
Methods and data: **Between January 1992 and December 2001, 201 patients with aortic coarctation were operated on at the Department of Cardiac Surgery of the Children's University Hospital, Bratislava. The three classes of aortic coarctation were represented: isolated coarctation, coarctation with ventricular septal defect (VSD) and coarctation with complex cardiac anomalies.**
Patients' preoperative, operative and immediate postoperative medical records were carefully studied with special attention paid to the type of lesion, patients' preoperative state, type of surgical technique employed, as well as the period of operation. For comparison, two equal time periods of follow-up were reviewed – 1992 to 1996 and 1997 to 2001. The overall postoperative conditions of patients were also regularly monitored.
Patient data were statistically analyzed using the JMP program version 4.04.
Results: **An overall survival of 90 % was recorded over the period of follow-up, ranging between one and ten years. A further break down showed a statistically significant difference between the various types of aortic coarctation, p=0.0001. Patients with simple or isolated coarctation had a survival rate of 100 %, those with ventricular septal defect (VSD) in addition to coarctation had a survival rate of 80 % while patients with associated complex cardiac anomalies had a survival rate of 65 %. An improvement on overall patient survival was recorded in the period between 1997 and 2001 – 96 % as against 86 % for the period between 1992 and 1996. On univariate statistical analysis, the following variables were identified as significant risk factors for death: 1) Complex cardiac anomalies (p<0.0001), 2) Age at operation less than one month (p<0.0001) and 3) Treatment prior to the year 1997 (p=0.02).**
Conclusion: **A considerable improvement on patient survival following surgery for coarctation of the aorta was recorded over the last five years. This could be attributed to new measures in preoperative, operative and postoperative care for patients with aortic coarctation.** ***(Tab. 4, Fig. 5, Ref. 8.)***
Key words: coarctation of the aorta.

Patient population/data distribution

A total of 201 pateints underwent surgery for coarctation of the aorta over a period of 10 years. 124 (62 %) of the operated patients were males, while 77 (38 %) were females, M:F=1.5. 139 (69.2 %) of all operated patients had simple or isolated coarctation (which includes coarctation with patent ductus arteriosus (PDA) and other minor cardiac anomalies). 35 patients (17.4 %) had coarctation with ventricular septal defects (VSD), while 27 (13.4 %) patients had coarctation with complex cardiac anomalies including hypoplastic left heart syndrome, transposition of the great arteries (TGA), Shone syndrome and others (see associated anomalies).

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Tab. 1. Yearly distribution of treated cases of aortic coarctation.

Year	No of pts	Isolated CoA	CoA with VSD	CoA with complex ICA
1992	28	22	5	1
1993	26	17	3	6
1994	25	20	2	3
1995	19	8	8	3
1996	26	15	6	5
1997	19	14	2	3
1998	18	15	2	1
1999	14	11	1	2
2000	16	9	5	2
2001	10	8	1	1
Total	201	139	35	27

Tab. 2. Age distribution of operated patients.

Age	No of pts	Percentage
1–28 days	65	32
28–180 days	41	20
6 months–2 years	15	8
2–6 years	28	14
6–18 years	52	26
Total	201	100

The yearly distribution according to type of aortic coarctation are shown on Table 1.

The mean age at operation was 3.5 years (range 2 days to 18 years). About 60 % of all operated patients underwent surgery before the age of 2 years while 74 % of all patients were operated on before the age of six years (Tab. 2). This is in line with our preferred surgical protocol, which favors correction of aortic coarctation as soon as it is diagnosed and the patient is deemed fit for surgery, mainly before or at about the age of 2 years.

The mean weight at operation was 13.8 kg, range: 1.7 to 67 kg. Expectedly, there was a close correlation between the age and weight at operation.

Associated anomalies

Some cardiac and non-cardiac anomalies are usually diagnosed in patients with aortic coarctation. Out of the 201 patients in our series, 135 (69 %) had at least one associated cardiac anomaly. The most common cardiac anomalies were various forms of valve lesions, occurring in about 44 % of cases. The most common valve anomaly was the bicuspid aortic valve, which occurred in 19 % of all cases of coarctation and in 43 % of all associated valve anomalies. We recorded 18 (9 %) cases of non-cardiac anomalies of which 4 (2 %) were cases of Turner's syndrome, 1 (0.5 %) case of Klippel Feil syndrome, 5 (2.6 %) cases of CNS lesions and 8 (4.4 %) cases of other anomalies ranging from gastrointestinal to metabolic disorders (Tab. 3).

Tab. 3. List of anomalies associated with coarctation of the aorta.

Associated anomalies	N*
a) Valve lesions	
- bicuspid aortic valve	37
- aortic regurgitation	4
- aortic stenosis	11
- mitral regurgitation	11
- mitral stenosis	3
- tricuspid regurgitation	15
- Shone syndrome	4
- Ebstein anomaly	1
b) Vascular anomalies	
- transposition of the great arteries	9
- a. lusoria	2
- partial anomalous pulmonary venous drainage (PAPVD)	1
- total anomalous pulmonary venous drainage (TAPVD)	1
c) Septal anomalies (excluding VSD)	
- secundum atrial defect/patent foramen ovale (PFO)	47
- atrioventricular septal defect (AVSD)	4
- single ventricle	2
d) Other complex cardiac anomalies	
- hypoplastic left heart syndrome (HLHS)	7
- double outlet right ventricle (DORV)	3
- double inlet left ventricle (DILV)	1

N* does not reflect the number of patients but anomalies; hence some patients had more than one associated anomaly.

Tab. 4. Causes of postoperative death.

Causes of postoperative death	N
Multiple organ failure/sepsis	5
Cardiac/respiratory failure	4
Metabolic disorder	1
Bronchopneumonia	2
Brain hemorrhage	1
Encephalomalacia	1

Surgical techniques

Four surgical techniques were mainly employed in the treatment of our patients:

Resection and end-to-end anastomosis (RETE)

Extended resection and end-to-end anastomosis (RETE-ext.)

Patch aortoplasty (Vosschulte) (PA)

Subclavian flap aortoplasty (Walddhausen) (SFAP)

A fifth surgical technique entailed total aortic arch reconstruction (TAR) using pericardial patch in some patients with associated aortic arch hypoplasia. On the whole, 101 patients (51 %) were treated by resection and end-to-end anastomosis, 24 patients (12 %) by extended resection and end-to-end anastomosis, another 24 patients by the subclavian flap angioplasty and 44 patients (22 %) by the patch aortoplasty technique. Total arch reconstruction was performed in 6 patients (3 %).

Since 1997, there has been an increased preference for the extended resection technique especially in neonates and infants (Fig. 1). The subclavian flap and patch aortoplasty technique are gradually being phased out. In fact, the subclavian flap tech-

Fig. 1. Extended resection and end-to-end anastomosis.

nique has not been used since 1997. This is due to the need to spare the subclavian artery, thus avoiding complications like the steal syndrome. The main reason, however, lies in the fact that the extended end-to-end technique is more physiological and as well enables extensive resection of the ductal tissue, thus decreasing the risk of recoarctation.

Patient survival and mortality

Patient survival over the period of follow-up was analyzed using the Kaplan-Meier technique. This technique shows the probability of survival (P) at a given point in time. Due to variation in time interval, the Kaplan-Meier estimate was preferred to the actuarial approach. Patients' survival according to such dependent variables as age at operation, type of coarctation, and year of operation was also determined.

An overall survival rate of 90 % was recorded over the period of follow-up (Fig. 2). The Kaplan-Meier plot in Figure 2 also shows that postoperative death mainly occurred within the first 6 months after surgery. These deaths mainly involved critically ill neonates with complex intracardiac anomalies in association with coarctation of the aorta.

Survival according to type of coarctation

Analysis of patients' survival according to type of coarctation showed significant differences among the groups (p=0.0001). Patients with isolated lesions (type 1) had the best chance of survival after operation, 100 % in our series. Patients with associated ventricular septal defects (type 2) had an 80 % survival rate over the period of follow-up. A survival rate of 65 % was recorded in patients who had complex intra-cardiac anomalies in association with coarctation of the aorta (type 3). A comparison of the survival rates for the various types of aortic coarctation is shown in Figure 3.

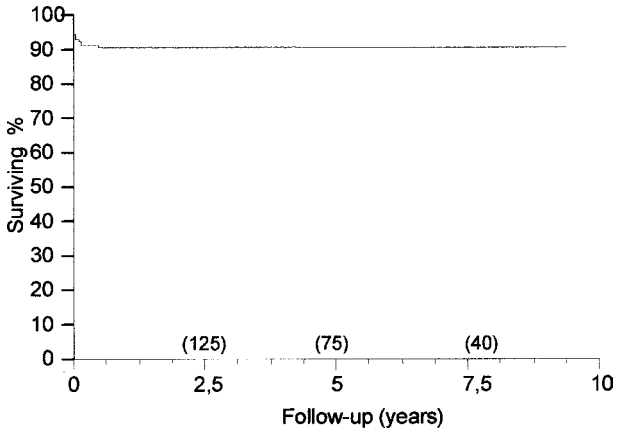


Fig. 2. Overall patient survival following surgical treatment of aortic coarctation. Patient at risk are in parenthesis.

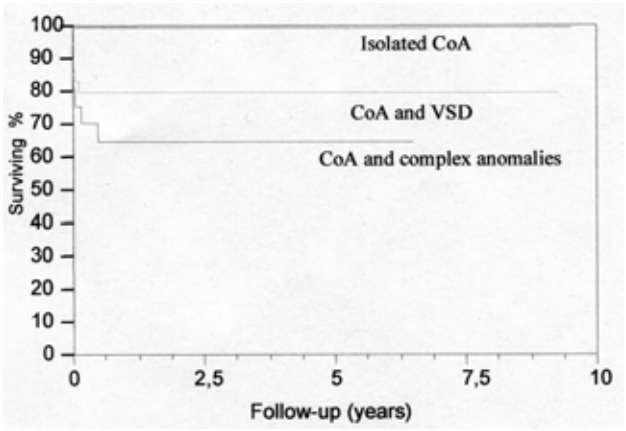


Fig. 3. Survival according to type of coarctation.

Survival according to year of operation

The graph in Figure 4 shows an improved overall survival of patients who underwent surgery in and after the year 1997 as compared to those operated on earlier (p=0.009). While there was an overall survival rate of 86 % before 1997, the situation was different in the period between 1997 and 2001, when we recorded an overall survival rate of 96 %. The rather dramatic improvement from 1997 to date can be attributed to the following factors:

a) Improved preoperative management of patients ensuring that critically ill neonates are referred to surgery in a stable condition.

b) Improved surgical technique and procedures such as the use of extended resection and end-to-end anastomosis, total arch reconstruction in patients with associated hypoplastic aortic arch, single-stage surgical treatment of patients with associated complex anomalies and the use of modified ultrafiltration following extracorporeal circulation.

c) Improved postoperative care and adequate follow-up of patients after discharge from hospital (Fig. 4).

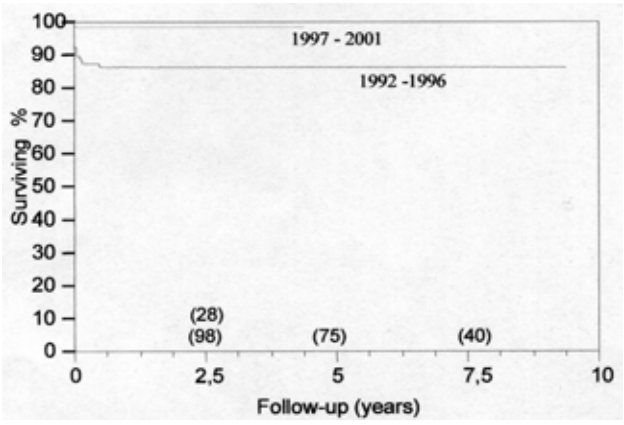
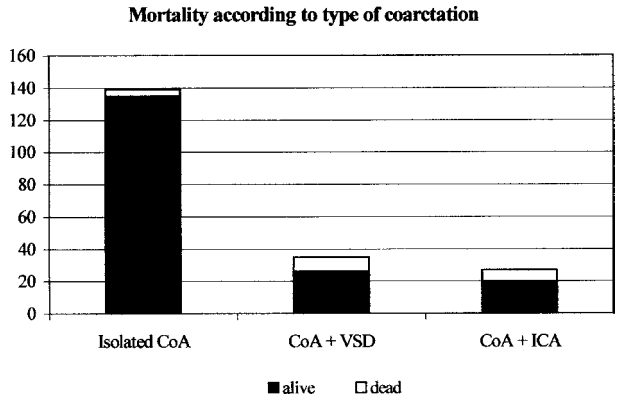


Fig. 4. Survival according to period of operation. Patients at risk are in parenthesis.



ig. 5. Mortality according to type of aortic coarctation.

Mortality

Among the 201 patients undergoing surgery, 20 (9.8 %) deaths were recorded. 4 (2 %) of the deaths were intra-operative while 16 (7.8 %) occurred after operation. All postoperative deaths occurred within the first month of surgery. The most common causes of postoperative death in our series were multi-organ failure due to sepsis and cardio-respiratory failures (Tab. 4).

Risk factors for death

On univariate analysis, the following factors were discovered to be in close correlation with the occurrence of death:

1) Coarctation with associated cardiac anomalies: Patients with aortic coarctation and other cardiac anomalies are more likely to die than are those with isolated lesions ($p<0.0001$). The relationship between these variables becomes even stronger with increasing complexity of the associated anomalies thus, the probability of dying decreases in this order: CoA+complex anomalies>CoA+VSD>Isolated CoA (Fig. 5).

2) Age at operation: There was a strong correlation between the age at operation and mortality ($p<0.0001$), with neonates and infants up to the age of six months accounting for over 95 % of all deaths. This however, had more to do with the fact that, patients with complex anomalies and signs of cardiac failure presented and were subsequenntly treated in neonatal age. Thus, age at operation per se cannot be said to be a risk factor for death. On multivariate analysis, $p=0.1$.

3) Operation before the year 1997: Neonates operated on before 1997 were at a greater risk of dying than those operated on after this period ($p=0.02$) – 17 (85 %) of the 20 deaths recorded over the period of our study occurred before 1997.

Discussion

Our study of patients undergoing surgery for coarctation of the aorta has shown an improved survival in the last five years. A review of the likely reasons for this improvement identified several measures involving preoperative, operative as well as postoperative management of patients.

The preoperative measures entail adequate care and monitoring of critically ill neonates with coarctation of the aorta in the cardiac intensive care unit (CICU). These involve the use of prostaglandins to keep the arterial ductus open and prevent imminent cardiac failure – a sign very common in neonates presenting with coarctation of the aorta. In addition, improved ventilation and adequate infection treatment have meant that most patients are now referred to surgery in a stable condition, reducing the number of cases of urgent surgical indication.

Changes in operative measures over the period in question include:

1) The use of extended resection and end-to-end anastomosis in neonates, which has been linked with a decrease in the rate of recoarctation in this age group and a consequent decrease in the need for re-intervention. In their study of 60 neonates treated by extended resection, Backer et al (1) recorded a recoarctation rate of 3.6 % within the first year of follow-up. In our unpublished data, the rate of recoarctation in neonates undergoing extended resection was 7.7 % over a mean follow-up period of 5 years as opposed to 32 % for simple resection and end-to-end anastomosis in the same age group, over the same period.

2) Total aortic arch reconstruction with the aid of a pericardial patch is proving to be an adequate treatment in patients with associated hypoplastia of the aortic arch. Even though this technique has been used in only a small number of patients in our institution, it has been associated with an improved survival and low rate of recoarctation.

3) Single stage treatment of patients with associated anomalies: This measure ensures total correction of coarctation of the aorta and all associated anomalies, thereby providing a complete solution to the patients' numerous hemodynamic problems. As in many other cases, adequate postoperative care is a guarantee for success.

4) Another surgery-related measures is the use of modified ultrafiltration (MUF) in patients following exra-corporeal circulation (ECC). The gains of modified ultrafiltration lie in effective reduction of total body fluid accumulated during ECC, increased hematocrit, reduced pulmonary resistance and thus, bet-

ter hemodynamics prior to weaning from ECC as well as a better post-operative course.

Postoperative patient management procedures in the CICU have also contributed to improved patient survival in our series. New ventilation possibilities as well as adequate inotropic support are some of the measures. Regular check-ups in the outpatient department has also ensured that patients with late postoperative complications are detected on time and referred to the appropriate unit for treatment.

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