Surgical management of Coarctation of the Aorta in the Pediatric Population: A fifteen-year experience

Kristi SALIAJ
Faculty of Medicine, Tirana, Albania
Corresponding author: Kristi Saliaj
E-mail: kristisaliaj@gmail.com

MD. Prof. Dr. Arben BABOCI
Department of Cardiovascular Surgery, University Hospital Center "Mother Teresa", Tirana, Albania

MD. Elvana RISTA
Department of Nephrology, Hygeia International Hospital, Tirana, Albania

MD. Dr. Sc. Altin VESHTI
Department of Cardiovascular Surgery, University Hospital Center "Mother Teresa", Tirana, Albania

MD. Dr. Sc. Saimir KUCI
Department of Cardiac Anesthesiology, University Hospital Center "Mother Teresa", Tirana, Albania

Introduction

Coarctation of the aorta is one of the most commonly encountered congenital heart defects (CHD), in the pediatric population. It is defined as a hemodynamically significant stenosis of the descending aorta, typically at the site of insertion of ductus arteriosus. It has an estimated incidence of 4-6 cases per 100,000 births, accounting for 5-8% of all congenital heart defects [1-4]. According to The New England Regional Infant Cardiac Program (NERICP), coarctation of the aorta is the fourth most common defect requiring surgery during the first year of life [5]. It can present as an isolated lesion or coexist with other cardiac or extra-cardiac congenital defects. The most common one include bicuspid aortic valve, patent ductus arteriosus, ventricular septal defect, mitral stenosis, berry aneurysms in the circle of Willis [6-8].

Clinical presentation is heterogenous, varying from asymptomatic to congestive heart failure, acute pulmonary edema and cardiogenic shock, depending on the degree of coarctation, the development of collateral circulation and age at presentation [9]. During the neonatal period patients may present with circulatory collapse and pulmonary edema, reduced or absent peripheral pulses, tachypnea, lethargy and progressive metabolic acidosis [9-13]. In early childhood, they present primarily with symptoms of congestive heart failure (CHF) including tachypnea, irritability, sweating, feeding difficulties and failure to thrive [9-13]. In adolescence and adulthood patients may be asymptomatic, presenting with non-specific symptoms including exercise intolerance, fatigue and cramps in the lower limbs, recurrent headaches, epistaxis, vertigo, tinnitus, high blood pressure and difference in blood pressure between arms and legs and reduced or absent peripheral pulses [9-13]. Treatment options for coarctation of the aorta include both surgery and catheter-based procedures. Several surgical techniques have been developed including extended resection with end-to-end anastomosis, prosthetic patch aortoplasty, subclavian patch aortoplasty, interposition grafting and extra-anatomic bypass grafting [9-11,13-18]. Catheter-based approaches include balloon angioplasty and stenting. The choice is based on the age at presentation, the type of coarctation and the presence of associated anomalies, as well as other patient-specific characteristics [9-11,18-23].

Methodology

We conducted an observational, retrospective, cohort study collecting data from the medical records of the Department of Cardiovascular Surgery at the University Hospital Center "Mother Theresa" Tirana (UHCMT), Albania. Baseline patient

characteristics, including demographic, clinical and surgical data of patients admitted in the span a fifteen year period (March 2004 – March 2020), were recorded. The aim of this study was the evaluation of the surgical management of coarctation of the aorta in the pediatric population, in our institution.

Results

In this single center study, a total of 85 patients were admitted with a diagnosis of coarctation of the aorta in the Department of Cardiovascular Surgery and underwent surgical treatment. Males represented 62.36% of our cohort.

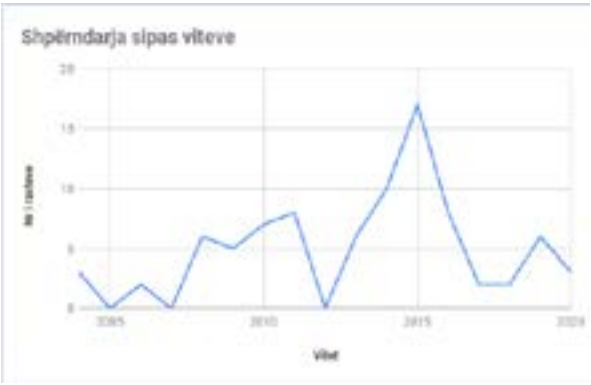
Based on their age distribution, patients were categorized in four groups (neonate, infant, early childhood, adolescents and adults). Adolescents and adults represented the subgroup with the highest number of patients 29.41% of the cohort, whereas patients in the early childhood, the subgroup with fewer patients, 17.65% of the cohort.

TABLE 1. Age distribution

	Nr of patients	Percentage
Neonate (0-28 days)	21	24.70%
Infant (29 days- <1 year old)	24	28.24%
Early childhood (1 year old -11 years old)	15	17.65%
Adolescents and adults (>12 years old)	25	29.41%
Total	85	100.0%

Looking at the yearly distribution, the highest incidence was reported in 2015 (17 patients) and the lowest in 2005, 2007 and 2012 (0 patients).

FIGURE 1. Yearly distribution

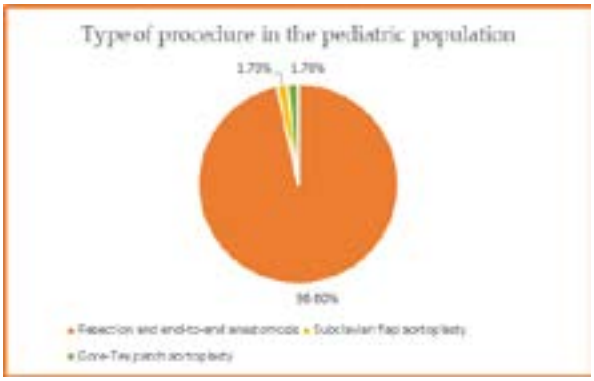


The pediatric population was arbitrarily defined as patients under the age of 12 years old, encompassing in total 60 patients. The age at presentation of the pediatric population ranged from 0.1 months (3 days) to 72 months (6 years old), the mean (SD) age was 5.96 ± 11.91.

20% of pediatric cohort presented as surgical emergencies, with acute pulmonary edema. 75% of the pediatric population (45 patients), presented with symptoms of congestive heart failure (CHF).

Our data revealed that resection with end-to-end anastomosis was the most frequently performed procedure in 96.6% of the pediatric cohort, followed by subclavian patch aortoplasty in 1.7% of patients and Gore-Tex patch aortoplasty in 1.7% of cases, as well.

FIGURE 2. Type of procedure in the pediatric population



Our study found that in a subgroup of patients, surgical treatment of coarctation of the aorta was associated with concomitant procedures, including patent ductus arteriosus (PDA) ligation in 19% of cases, pulmonary artery banding in 3.4%

of cases and subaortic membrane resection in 1.6% of cases. In 76% of patients coarctation repair was performed alone.

In a subgroup of patients, a second concurrent heart defect was present. Of these associated congenital heart defects, patent ductus arteriosus (PDA) was the most common one, accounting for 70.6 % of cases, followed by ventricular septal defect (VSD) in 17.6% of cases and subaortic stenosis and ascending aorta aneurysm in 5.9% of cases, respectively.

Table 2. Associated congenital heart defects repaired concomitantly

Associated anomalies repaired concomitantly	Nr of patients	Percentage
PDA	12	70.60%
VSD	3	17.60%
Subaortic stenosis	1	5.90%
Ascending aorta aneurysm	1	5.90%

All patients (100%) were found to be hemodynamically stable at the end of surgery. All surgical complications developed in the post-operative period. Mean (SD) clamping time was estimated to be 20.68 ± 10.16 min.

Post-operative complications including chylothorax (50%), acute kidney injury (25%) and anastomotic thrombosis (25%), developed in 6.7% of the pediatric population.

Early post-operative complications	Nr of cases	Percentage
Chylothorax	2	50%
Acute kidney injury (AKI)	1	25%
Anastomotic thrombosis	1	25%

Mortality in our cohort was 5%, with 3 patients dying due to complications in the post-operative period.

Discussion

Coarctation of the aorta is one of the most common congenital heart defects and patients can benefit from a broad spectrum of available surgical treatment options.

In our cohort, there was a slight male predominance. 62% of the cohort were male, with a male/female ratio of approximately 1.6/1. These findings are consistent with reported statistics [5]. The highest incidence was reported in 2015

(17 patients) and the lowest in 2005, 2007 and 2012 (0 patients). The mean (SD) age at presentation was 5.96 ± 11.91 months.

In our study, 20% of the pediatric population presented as surgical emergencies with acute pulmonary edema and underwent surgical repair within the first two weeks of the neonatal period. The predominant clinical presentation in the pediatric population was congestive heart failure (CHF), in 75% of all patients, consistent with current literature on clinical presentation of coarctation of the aorta with signs of heart failure in the first year of life [9-13].

In our study, the mainstay surgical procedure in the pediatric population was resection with end-to-end anastomosis (96.6%), followed by subclavian patch aortoplasty (1.7%) and Gore-Tex aortoplasty (1.7%). These findings are consistent with current literature and recommended practices [9-11,13-18]. Extended resection with end-to-end anastomosis is the preferred surgical techniques in native, discrete coarctation in infancy and early childhood [9-11,13-18,24,25]. It allows the complete resection of the stenotic segment without the use of any prosthetic material and is associated with low mortality and recoarctation rates [11,24,25].

Prosthetic patch aortoplasty offers several advantages including the treatment of longer coarctation segments and lower recoarctation rates, however it is associated with a higher prevalence of aortic aneurysms [9-11,26,27]. Despite the use of PTFE prosthetic patches instead of Dacron ones, the risk for aortic aneurysms remains higher, compared to other techniques [9-11,26,27]. Subclavian flap aortoplasty can also be used to repair longer segments of coarctation, but it can lead to the development of the subclavian steal syndrome [9-11,28].

Interposition grafting and extra-anatomic bypass grafting are usually employed in the treatment of long hypoplastic aortic segment and in adult patients, as prosthetic grafts pose growth limitations in the pediatric population [11,29,30].

Catheter-based treatments including balloon angioplasty and stenting are less preferred in infants and young children, as they are associated with higher rates of restenosis and late aneurysm formation compared to surgical repair [9-11,18-23,31-33]. Balloon angioplasty and stenting are primarily used in the treatment of native coarctation in older children and adults, as well as in recurrent coarctation [9-11,18-23,31-33]. Balloon angioplasty can also be used as a palliative procedure in critically ill neonates, providing some clinical improvement until they are stable to undergo surgical repair [33].

In our cohort in a subgroup of patients, surgical treatment of coarctation of the aorta was associated with concomitant procedures, including patent ductus arteriosus (PDA) ligation in 19% of cases, pulmonary artery banding in 3.4% of cases and subaortic membrane resection in 1.6% of cases.

Our study found that the most common concomitant congenital heart defect to

be repaired in tandem, was patent ductus arteriosus (PDA) accounting for 70.6% of cases, followed by ventricular septal defect (VSD) in 17.6%, subaortic stenosis and aneurysm of the ascending aorta in 5.9% of cases, respectively.

Complications of coarctation of the aorta can be classified as early and late complications. Early complications are associated with the post-operative period and they include paradoxal hypertension, chylothorax and paraplegia [9]. In our cohort, the most common one was chylothorax (50%), followed by acute kidney injury (25%) and anastomotic thrombosis (25%), developing 6.7% of the pediatric population.

Late complications are associated with the type of surgical procedure and the pathogenesis of coarctation and they influence the long-term morbidity and mortality rates in this population. These complications include aneurysm formation, coronary heart disease, arterial hypertension, cerebrovascular complications and recurrent coarctation [9,34-39].

Following surgery, in the early post-operative period all patients (60 patients) were hemodinamically stable. The mortality rate in our cohort was 5%, consistent with current studies and literature, emphasizing that surgical repair of coarctation of the aorta is safe and effective [11].

Conclusion

Despite its reported prevalence, coarctation of the aorta among pediatric patients remains sporadic in our population. Our study revealed that early surgical repair is safe and effective, associated with excellent outcomes and low mortality rates, consistent with current literature and recommended practices. The challenges in the management of coarctation of the aorta in the pediatric population don't lie in the treatment of the stenotic segment, but in preventing long-term cardiovascular complications. Risk stratification of patients and long-term follow-up programs can improve outcomes and survival.

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