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Results of surgical treatment of aortic coarctation

Resultados del tratamiento quirúrgico de la coartación aórtica

Dr. C. Alfredo M. Naranjo Ugalde¹ D,Dr. C. Eugenio Selman-Housein Sosa¹ Dr. Gilberto Bermúdez Gutiérrez¹ Dra. Lais A. Ceruto Ortíz¹ Dr. Alexander González Guillén¹

¹ Department of Cardiovascular Surgery. Cardiocentro Pediátrico "William Soler"

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Corresponding author:

PhD, MD, Alfredo M. Naranjo Ugalde naranjob@infomed.sld.cu

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ABSTRACT:

Introduction: Aortic coarctation is defined as the congenital narrowing of this artery that causes partial obstruction to blood flow. Its clinical presentation depends on the severity of the injury and the associated malformations. It can cause high blood pressure, heart failure, and pulmonary hypertension. **Objectives:** Describe the results of the surgical treatment of aortic coarctation. **Methodology:** A descriptive cross-sectional study was carried out on 188 patients undergoing surgery for aortic coarctation from 2010 to 2023 at the William Soler Pediatric Cardiocenter, a national reference center for the treatment of this condition. Results: Of the 188 patients operated on, 62.7% were vounger than three months, with a predominance of males. Although the majority suffered from isolated aortic coarctation, a high percentage was associated with other cardiac anomalies and required an associated pulmonary cerclage-type procedure 27.7% were performed urgently. The most used surgical technique was coartectomy with end-to-end anastomosis, although depending on age and associated anomalies, others were perfor med. Mortality was 6.9% and was related to age weight and surgical priority. Conclusions: The surgical treatment ofb aortic coarctation is a safe procedure at the William Soler Pediatric Cardiocenter, its mortality is not high and is not related to the surgical technique performed.

Keywords: Aortic coarctation, congenital heart disease, cardiac surgery RESUMEN

Introducción: La coartación aórtica se define como el estrechamiento congénito de esta arteria que causa una obstrucción parcial al flujo sanguíneo. Su forma de presentación clínica depende de la severidad de la lesión y de las malformaciones asociadas. Puede provocar hipertensión arterial, insuficiencia cardiaca, e hipertensión pulmonar. Objetivos: Describir los resultados del tratamiento quirúrgico de la coartación aórtica. Metodología: Se realizó un estudio descriptivo de corte transversal en 188 pacientes intervenidos quirúrgicamente de coartación aórtica desde el 2010 hasta el 2023 en el Cardiocentro Pediátrico William Soler, centro de referencia nacional para el tratamiento de esta afección. Resultados: De los 188 pacientes operados, el 62,7 % fueron menores de tres meses, con predominio del sexo masculino. Aunque la mayoría padecían coartación aórtica aislada, un elevado porcentaje se asoció a otras anomalías cardiacas y requirió procedimiento asociado tipo cerclaje pulmonar. El 27,7 % se practicó de manera urgente. La técnica quirúrgica más utilizada fue la coartectomía con anastomosis término-terminal, aunque en dependencia de la edad y las anomalías asociadas, se ejecutaron otras. La mortalidad fue de 6,9 % y presentó relación con la edad, el peso y la prioridad quirúrgica. Conclusiones: El tratamiento quirúrgico de la coartación aórtica es un procedimiento

seguro en el Cardiocentro Pediátrico William Soler, su mortalidad no es elevada y no se relaciona con la técnica quirúrgica realizada.

Palabras clave: Coartación aórtica, cardiopatías congénitas, cirugía cardiaca

INTRODUCTION

Aortic coarctation is defined as a congenital narrowing of the aorta at any level beyond the aortic arch, causing partial obstruction of distal blood flow and resulting in a difference between blood pressure measured in the upper limbs compared to the lower limbs. It occurs most frequently at the aortic isthmus and in the periductal region, either proximal or distal to the ductus arteriosus or its remnant, the ligamentum arteriosum.¹

It accounts for approximately six to eight percent of all congenital heart diseases^{2,3}, with an incidence rate of three to four per 10 000 live births and a predominance in males.^{1,4,5}

First described by Morgagni in 1760, it is the congenital malformation most frequently diagnosed in newborns and infants.² Symptom onset in these age groups is related to the physiological closure of the ductus arteriosus and, occasionally, to some degree of aortic underdevelopment, depending on the severity of the narrowing.^{6,7}

Mild forms may be diagnosed at any age due to the development of systemic high blood pressure and progressive left ventricular dysfunction. In more severe cases, heart failure and pulmonary hypertension allow for diagnosis in early life (newborns and infants).⁸

Anatomically, the lesion may be localized, presenting as a fibrous ring within the arterial lumen with a central or eccentric orifice, distal to the origin of the left subclavian artery (juxtaductal), or as an elongated narrow segment involving the aortic isthmus (the segment between the left subclavian artery and the ductus arteriosus), and may be associated with hypoplasia of the transverse aortic arch (the segment between the origins of the left carotid and left subclavian arteries) in 20% of cases. Other commonly associated anomalies include bicuspid aortic valve, ventricular septal defect, mitral valve anomalies, or subaortic stenosis. ^{1,5}

The natural course of aortic coarctation, characterized by high blood pressure, may lead to serious complications such as aortic dissection, heart failure, or rupture of a cerebral aneurysm.^{9,10}

The first surgical correction, consisting of resection of the coarcted segment (coartectomy) with end-to-end anastomosis, was reported by Clarence Crafoord in 1944. Subsequently, surgical variants have been described using the left subclavian artery or prosthetic material for aortoplasty. Endovascular therapy has also contributed to managing this condition by dilating

the coarcted area, with or without stent placement. 12,13 The presence of structural cardiac lesions requires combined treatment during the same surgical procedure, including interventions such as the closure of ventricular septal defects, pulmonary artery banding, or valve repairs. 8,14

Since 1986, the *Cardiocentro Pediátrico "William Soler"* has served as the national reference center for the treatment of congenital heart disease in Cuba. The aim of this study was to describe the outcomes of surgical treatment for aortic coarctation, with a particular focus on hospital mortality and its associated factors.

METHOD

A descriptive, cross-sectional study was conducted, including all patients who underwent surgical repair of aortic coarctation at the Cardiocentro Pediátrico "William Soler" from January 2010 to May 2023.

Among the associated lesions, only those that, from a surgical perspective, determined the choice of surgery technique were considered. The persistence of patent ductus arteriosus was excluded as it was obligatorily ligated or divided during the definitive surgical procedure in all cases.

Perioperative data were retrieved from the DELFOS database¹⁵ of the Department of Cardiovascular Surgery.

All surgeries were performed in accordance with established guidelines for these interventions and by the same surgical team. A left posterolateral thoracotomy was employed as the surgical approach in all patients. ¹⁶

Study variables were expressed according to their respective summary measures: quantitative variables are presented as mean and standard deviation, and qualitative variables as counts and percentages. Associations with hospital mortality were assessed using the chi-square test of independence, Fisher's exact test, or Welch's t-test for unequal variances, with p<0.05 considered statistically significant.

The study adhered to the ethical principles outlined in the Declaration of Helsinki.¹⁷

RESULTS

From October 2010 to March 2023, a total of 188 surgical procedures for aortic coarctation were performed at the Cardiocentro Pediátrico "William Soler", representing 5.85% of all surgical procedures during that period.

Table 1 presents the association between hospital mortality, defined as state at discharge deceased, and

a series of preoperative variables. Hospital mortality occurred in 13 patients (6.9%) and was significantly associated with age, weight, and surgical priority. Patients aged 1 to 3 months comprised the largest group (48.9%), while newborns accounted for 13.8%. Urgent procedures were performed in 27.7% of cases, including patients with higher clinical risk, particularly newborns.

No significant association was found between hospital mortality and sex or type of diagnosis. The majority

of patients were male (62.2%). Isolated aortic coarctation was the most frequent diagnosis (76.1%). The remaining patients diagnosed with coarctation associated with major cardiac anomalies, such as ventricular septal defect, aberrant right subclavian artery, transposition of the great arteries, or single ventricle, required more complex surgical techniques. Among 12 patients undergoing repair for recoarctation, a technically challenging procedure, no deaths were reported.

Table 1. Characteristics of the patients operated on aortic coarctation. *Cardiocentro Pediátrico "William Soler.*

Characteristics	Global (%) n = 188	State at d					
		Alive (%)	Deceased (%)	p-value			
		n=175 (93,1)	n=13 (6,9)				
Age				<0,001*			
Newborn	26 (13,8)	20 (76,9)	6 (23,1)				
1 to 3 months	92 (48,9)	85 (92,4)	7 (7,6)				
4 to 11 months	35 (18,6)	35 (100)	0 (0,0)				
≥ 1 year	35 (18,6)	35 (100)	0 (0,0)				
Sex				0,059*			
Male	117 (62,2)	108 (92,3)	9 (7,7)				
Female	71 (37,8)	67 (94,3)	4 (5,7)				
Mean weight in kg ± SD	$24,5 \pm 16,8$	24,6 ± 16, 8	$2,6 \pm 0,36$	<0,001†			
Diagnosis				0,383*			
Isolated CoAo	143 (76,1)	134 (93,7)	9 (6,3)				
CoAo-VSD	24 (12,8)	21 (87,5)	3 (12,5)				
Recoarctation	12 (6,4)	12 (100)	0 (0,0)				
CoAo-ARSA	5 (2,6)	5 (100)	0 (0,0)				
CoAo-TGA or SV	4 (2,1)	3 (75)	1 (25)				
Surgical priority				<0,001*			
Elective	136 (72,3)	132 (97,1)	4 (2,9)				
Urgent	52 (27,7)	43 (82,7)	9 (17,3)				
*Chi-square test of independence: Fisher's exact test							

^{*}Chi-square test of independence; Fisher's exact test

ARSA: Aberrant Right Subclavian Artery; CoAo: Coarctation of the Aortic; SD: Standard Deviation; SV: Single Ventricle;

TGA: Transportation of Great Arteries; VSD: Ventricular Septal Defect

The surgical techniques applied across different age groups are shown in figure 1. Regarding surgical techniques, coartectomy with end-to-end anastomosis was the most frequently performed procedure (67 patients) across all age groups, followed by extended end-to-end anastomosis in 53 children under one year. Coartectomy with Waldhausen technique was performed in 32 patients across all ages. Patch aortoplasty was performed in 28 pediatric patients, predominantly those older than one year. Coartectomy with end-to-end anastomosis plus superior subclavian flap was performed in three patients, including one newborn and two infants aged 1-3 months.

Out of the eight surgical techniques listed in figure 1, six were performed in the newborn group and five in the one- to three-month group, which is the largest group in the series. Patients aged one year or older received three of the surgical techniques presented.

Figure 2 illustrates the frequency of any coarctectomy procedure performed in conjunction with pulmonary artery banding. This combination was necessary due to the presence of a ventricular septal defect (VSD) in 27 patients aimed at preventing or delaying pulmonary hypertension caused by isolated VSD or transposition of the great arteries (TGA). It is notable that the procedure was required in all age groups, although

[†]Welch's t-test for unequal variances

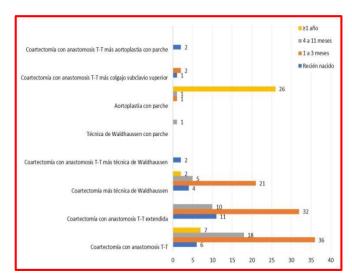


Figure 1 - Surgical techniques employed in different age group

Legend: T-T: Término-Terminal (End-to-end)

the majority were in newborns and infants aged one to three months, with 9 and 12 cases respectively.

The combination of procedures may be linked to higher mortality. Table 2 shows that the use of any coarctectomy technique combined with the pulmonary artery banding procedure did not increase hospital mortality across the studied age groups.

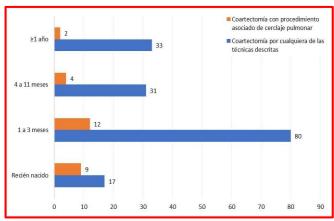


Figure 2 - Association of any coarctectomy technique with pulmonary artery banding procedure in different age groups

Among newborns undergoing coarctectomy with the associated pulmonary artery banding procedure, there was only one death, representing 11.1%, compared to 29.4% mortality in those who did not undergo combined procedures, showing statistically significant differences. In the one- to three-month age group, the combination of procedures resulted in a hospital mortality rate of 16.6%, while those without the combined procedure had a mortality of 6.2%.

The postoperative complications experienced by patients according to their age are shown in figure 3. As seen, newborns and infants between one and three

months experienced all the complications listed, except for bleeding, which only occurred in children aged one year or older. Respiratory complications, such as pneumothorax, phrenic nerve paralysis, subcutaneous emphysema, and prolonged mechanical ventilation, were more common. The latter occurred exclusively in those under three months of age, along with severe sepsis and ventricular dysfunction. Immediate recoarctation appeared only in newborns.

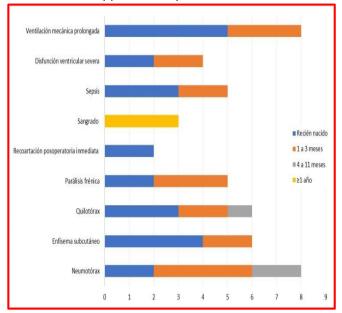


Figura 3 - Postoperative complications by age

DISCUSSION

Surgical treatment for aortic coarctation is commonly performed at nearly all pediatric cardiovascular surgery centers. Despite the availability of percutaneous treatments for many patients, surgery remains an important therapeutic option, especially in cases with associated anomalies or the presence of a hypoplastic aortic arch. A Spanish report compiling the experience from an average of 55 surgical centers over four years, states that 4.5% of congenital heart disease surgeries were for aortic coarctation, a figure consistent with the current series.¹⁸

The predominance of male patients is consistent in agreement with findings from various However, it should not be assumed as a pattern for this congenital anomaly, as some authors have reported a predominance of female patients or an even gender distribution.^{22,23}

In the present report, nearly 20% of the patients undergoing surgery had an associated VSD, whereas Oliveira et al.¹⁹ report only 9%. This led to the use of of combined coarctectomy techniques with pulmonary artery banding, which was not reported by these authors. Neither a bicuspid aorta nor a persistent ductus arteriosus was recorded as an associated lesion, as the surgical technique was not determined. Some authors do not consider a bicuspid aorta a

Table 2. State at discharge by age and differences in hospital mortality between isolated coarctectomy and coarctectomy combined with pulmonary artery banding procedure

	State at discharge by age				
Combination of procedures	Newborn n=26		One to three months age n=92		
	Alive(%)	Deseased (%)	Alive(%)	Deseased (%)	
Coarctectomy by any of the techniques presented (Isolated Coarctectomy)	12 (70,6)	5 (29,4)	75 (93,8)	5 (6,2)	
Coarctectomy combined with pulmo- nary artery banding procedure	8 (88,9)	1 (11,1)	10 (83,4)	2 (16,6)	
Total	20 (76,9)	6 (23,1)	85 (92,4)	7 (7,6)	

congenital malformation, but rather an anatomical variant of importance in the presentation of aortic stenosis.⁶

Early treatment of coarctation guarantees better quality of life and life expectancy. As noted in the present report, most patients undergoing surgery during the infant period constitute the largest patient group in several reports. 14,19,20,23,24 Although technological advances allow for surgical treatment at very early ages, it has also been reported that early treatment may favor the subsequent recoarctation. 8,25 In the current experience, only 1.3% of patients under one year of age underwent recoarctation during the analyzed decade.

The highest percentage of patients who underwent recoarctation had received percutaneous treatment as the first line of therapy, similar to some references found.^{26,27}

The incidence of recoarctation was observed to be associated in several cases with hypoplasia of the aortic arch and the presence of an aberrant right subclavian artery, as noted by other authors. 10,28

As reported in other studies, the most frequently used surgical technique in infants under one year was co-arctectomy with end-to-end anastomosis or extended anastomosis in cases of hypoplastic aortic arch. 19,23,29,30 In the authors' experience, combining coarctectomy and anastomosis with left subclavian artery descent (Waldhausen technique) or patch augmentation significantly reduces the chances of recoarctation in the medium to long term when the patient is under three months of age. This is because its incidence has been associated with the presence of residual periductal tissue, and these technical variations effectively exclude this possibility. 31,32

In children over one year old, the aortic diameter allows for preservation of the periductal tissue and release of the obstruction by enlarging the vessel's anterior face. Due to the frequent presence of collateral arteries, patch aortoplasty was the preferred technique. Both this and the Waldhausen technique provide the surgeon with a lower possibility of collateral

injury as they do not involve manipulation of the posterior aortic surface.³¹

Some reports do not find a relationship between hypoplastic aortic arch and being a very young or low-weight newborn. However, other authors observe some degree of relationship between these factors. The stated that once the coarcted area of the aorta is released in a newborn, the aortic arch continues to grow, potentially leading to a non-hypoplastic arch, complicating further correlation with other variables.

Immediate complications mainly occurred in the youngest group and were related to the surgical access procedure, such as the thoracotomy, rather than manipulation of the aorta itself. A similar experience is reported by other authors, who ensure good outcomes with patch aortoplasty procedure, consistent with our findings.²⁰

Prolonged mechanical ventilation due to respiratory causes, severe ventricular dysfunction with poor therapeutic response, and severe sepsis were more frequent in the younger age group, due to the severity of the aortic coarctation and its consequences. Brazilian authors mention ventricular failure, arrhythmias, and neurological damage as the most frequent complications.¹⁹

Paraplegia, a feared complication, reported by some authors²⁰, was not observed in this series. The use of mild hypothermia, as well as the systematization of the procedure, has contributed to this outcome.¹⁶

The hospital mortality found is similar to the Brazilian report¹⁹, although higher than reported in other series.^{21,36} Some authors highlight the reduction in hospital mortality when compared to earlier decades, attributing it to the experience of the surgical team, patient volume, and the continuous improvement of care protocols.³⁸⁻⁴⁰ This view aligns with the findings of the current research team.

Age is identified as a statistically significant factor associated with surgical mortality. Notably, no deaths occurred in patients older than four months.

In newborns, cardiovascular decompensation occurs very early, with ventricular dilation and endocardial elastosis contributing to poorer postoperative recovery. Some hospitals report that up to 50% of infants operated on before three months of age had received prior percutaneous treatment as newborns, which, in the authors' opinion, might reduce the frequency of postoperative complications. However, it would be worth considering the risks of anesthesia and catheterization at that age and the high rate of recoarctation following this procedure. ^{5,20}

Hospital mortality was lower in newborns with VSD associated with aortic coarctation than in those who underwent both coarctectomy and pulmonary artery banding. This may seem counterintuitive, but the existence of a non-restrictive VSD delays the negative effects of aortic coarctation on the left ventricle, allowing pathophysiological blood flow through the VSD.

However, in infants between one and three months of age, the opposite was observed, the mortality was higher in the group requiring the associated pulmonary artery banding procedure. At this age, there is greater volume overload and reduced pulmonary resistance. Increased pulmonary flow with tissue damage and pre-existing left ventricular dilation may have contributed to this outcome from a pathophysiological perspective.

Some researchers point out that the immaturity of the newborn and the onset of the disease with heart failure influence the postoperative evolution. This latter condition is often the driving force for urgent surgery, identified as a risk factor. However, it is also stated that age or weight below 2.5 kg do not currently correlate with mortality. 41

Regarding the results of coarctectomy associated with septal defects or persistent ductus arteriosus, conditions with increased pulmonary pressures, and the use of pulmonary artery banding in the former, some authors report no significant difference in both age groups. The balance between pulmonary and systemic flow is critical, partially ensured by elevated pulmonary pressures in the first month of life.⁴²⁻⁴⁴

However, other studies identify age under three months as a factor associated with poorer surgical outcomes, attributed to clinical deterioration present at the time of surgery as a result of heart failure. 45,46

CONCLUSIONS

The results of surgical treatment for aortic coarctation are satisfactory and the procedure is considered safe. Surgical mortality was not high in the presented series and was associated with age, weight, and surgical priority. The older age groups showed no mortality. Neither the diagnosis nor the surgical technique was linked to mortality, even when combined with pulmonary artery banding. Complications were more frequent in younger age groups.

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AUTHORS' CONTRIBUTION:

Conceptualization: Alfredo Mario Naranjo Ugalde Data curation: Alfredo Mario Naranjo Ugalde, Lais Angélica Ceruto Ortiz, Alexander González Guillén Formal analysis: Alfredo Mario Naranjo Ugalde Funding acquisition: Eugenio Selman-Housein Sosa Research: Alfredo Mario Naranjo Ugalde, Eugenio Selman-Housein Sosa, Gilberto Bermúdez Gutiérrez, Lais Angélica Ceruto Ortiz, Alexander González Guillén Methodology: Alfredo Mario Naranjo Ugalde, Eugenio Selman-Housein Sosa, Gilberto Bermúdez Gutiérrez, Lais Angélica Ceruto Ortiz, Alexander González Guillén Project administration: Alfredo Mario Naranjo Ugalde, Eugenio Selman-Housein Sosa

Resources: Provided by the department Software: Lais Angélica Ceruto Ortiz

Supervision: Alfredo Mario Naranjo Ugalde, Eugenio

Selman-Housein Sosa

Validation: Alfredo Mario Naranjo Ugalde, Eugenio

Selman-Housein Sosa

Visualization: Alfredo Mario Naranjo Ugalde, Lais Angélica Ceruto Ortiz

Writing-original draft: Alfredo Mario Naranjo Ugalde, Lais Angélica Ceruto Ortiz

Writing-review and editing: Alfredo Mario Naranjo Ugalde, Lais Angélica Ceruto Ortiz, Eugenio Selman-Housein Sosa

