ORIGINAL ARTICLE

Vol. 11 No. 1 January-March 2026

doi: 10.35366/122229

Prevalence of aortic arch defects in the neonatal period in a reference hospital. Are we achieving optimal results?

Prevalencia de defectos de arco aórtico en el periodo neonatal en un hospital de referencia. ¿Estamos logrando resultados óptimos?

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ABSTRACT

Introduction: coarctation of the aorta and interruption of the aortic arch are frequent pathologies in the neonatal stage, being one of the main causes of cardiogenic shock and death. **Objective:** the objective of this study is to know the frequency, demographic data, management, and surgical results of aortic arch defects in the neonatal period in a period of five years. Material: this is a descriptive, observational, cross-sectional, retrospective, and case series study, from 2019 to 2024. Recording of media, maximum, and minimum was made for continuous numerical variables and for categorical variables the percentage was measured. **Results:** we found a total of 44 patients with predominance in the male gender (56.8%), 11.4% of the cases represent Interruption of the Aortic Arch, the maximum transfer time for management was 18 days in 9.1% of the cases, mortality overall was 13.6%. **Conclusions:** it is common for aortic arch defects in the neonatal period to present as critical heart disease, that is, pathologies that if not detected and treated in time, patients have a high risk of dying in the first ten days of life. It is important to perform an adequate physical examination and cardiological screening for the timely detection and transfer of these patients.

Keywords: aortic coarctation, congenital heart disease, cardiac surgery.

RESUMEN

Introducción: *la coartación de la aorta y la interrupción de arco aórtico son patologías frecuentes en la etapa neonatal siendo una de las principales causas de choque cardiogénico y muerte.* ***Objetivo:*** *el objetivo de este estudio es conocer la frecuencia, datos demográficos, manejo y resultados quirúrgicos de los defectos de arco aórtico en el periodo neonatal en un periodo de cinco años. Material: se trata de un estudio descriptivo, observacional, transversal, retrospectivo y serie de casos, del 2019 a 2024. Se realizó registro de medias, máximos y mínimos para las variables numéricas continuas y para las variables categóricas se midió el porcentaje.* ***Resultados:*** *encontramos un total de 44 pacientes con predominio en el género masculino (56.8%), el 11.4% de los casos representan interrupción de arco aórtico, el tiempo máximo de traslado para manejo fue de 18 días en el 9.1% de los casos, la mortalidad global fue de 13.6%.* ***Conclusiones:*** *los defectos de arco aórtico en el periodo neonatal es común que se presenten como cardiopatías críticas, es decir patologías que si no se detectan y tratan a tiempo los pacientes tienen un alto riesgo de fallecer en los primeros diez días de vida. Es importante realizar una exploración física adecuada y el tamiz cardiológico para la detección y traslado oportuno de estos pacientes.*

Palabras clave: *coartación aórtica, cardiopatía congénita, cirugía cardíaca.*

How to cite: Massini-Aguilera I, López-Taylor J, Medina-Andrade MA, Ramírez-Cedillo D, Jiménez-Fernández CA, Ramírez FC, et al. Prevalence of aortic arch defects in the neonatal period in a reference hospital. Are we achieving optimal results?. Cir Card Mex. 2026; 11 (1): 9-12. https://dx.doi.org/10.35366/122229

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Received: 08-07-2025. Accepted: 13-09-2025.

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Abbreviation:
IAA = interrupted aortic arch

Aortic arch defects are congenital anomalies that frequently manifest with signs of cardiogenic shock during the neonatal period.[1] Coarctation of the aorta accounts for approximately 5 to 7% of all congenital heart defects, with a prevalence of 0.2 per 1,000 live births, occurring predominantly in males.[2,3] Interrupted aortic arch (IAA), characterized by a complete discontinuity between the ascending and descending aorta, has a reported prevalence of 0.03 per 1,000 live births.[4] In 1959, Celoria and Paton introduced a classification system based on the site of interruption relative to the origins of the major branches, dividing IAA into types A, B, and C.[5] During the neonatal stage, these conditions commonly present as critical congenital heart defects, specifically as duct-dependent lesions, whose clinical presentation may mimic neonatal sepsis.[5]

In our state, only a few public hospitals perform cardiac surgery, which frequently leads to significant delays in care for these patients, substantially increasing their mortality.[6]

The aim of this study is to evaluate the surgical outcomes of neonates with aortic arch defects treated at our institution and, indirectly, to assess the time required for their transfer to a surgical center.

MATERIAL AND METHODS

This was a descriptive, observational cross sectional and retrospective study of patients who underwent aortic arch defect repair during the neonatal period at our institution. The study was conducted over a five-year period spanning from 2019 to 2024.

Demographic data, surgical outcome, and postoperative follow up were described. Mean, maximum and minimum values were recorded for continuous numerical variables, and percentages were measured for categorical variables.

Ethical considerations: informed consent was obtained from the patients 'families for the study, and the study was approved by the hospital's ethics committee.

RESULTS

A total of 44 patients were admitted, 56.8% of whom were male. Eleven percent had interrupted aortic arches one of which was type C and two were types A and B, respectively. Twenty-three patients with aortic coarctation were accompanied by other heart conditions, primarily atrial septal defects, ventricular septal defects and bicuspid aorta; three patients had imperforate anus, two had Turner syndrome and three patients with interrupted aortic arch had dysmorphic syndrome.

The mean weight was 3.2 kg with a mean maternal age of 23 years and a mean gestational age of 37 weeks. Demographic data are presented in *Table 1*.

The mean transfer time in days was 12 (P25: 9, P75: 12). Patients were divided into two group gassed on the time: less than 10 days and equal to or greater than 10 days. The first group was found to have only 32.6% of the patients transferred while group 2 accounted for 67.4%. All of the patients with interrupted aortic arch were in the greater than 10 days group.

The mean age at surgery was 15 days (P25: 8, P75: 20). Patients were divided into two groups was 10 days or younger, representing 28.2%, and the second group was 10 days or older, representing 71.7%. The mean weight in kilograms at the time of surgery was 3.2 (P25: 3, P75: 3.5). Regarding clinical presentation 61.4% (N = 27) presented cardiogenic shock.

The preoperative echocardiogram showed a mean gradient of the descending aorta in mmHg of 54 (P25: 45, P75: 65). The left ventricular function measured in percentage was 55 (P25: 43, P75: 67). The mean z scores include aortic valve 1.4, transverse arch 1.8, aortic arch −1.6, coarctation zone −3, and descending aorta 1.9. Echocardiographic values are listed in *Table 2*.

The most common surgical procedure performed was extended end to end cortectomy (50%), and graft placement was the least common (9.1%). However, it was the primary surgical technique for repairing cases with interrupted aortic arch. The mean aortic cross clamping time was 18 minutes, and the surgical time was 161 minutes. All surgical date is presented in *Table 2*. Median sternotomy was performed in patients with interrupted aortic arch and in those with severe aortic arch hypoplasia, which accounted for 16% of patients.

Table 1: Demographic data of the study population.

Variable	Mean [range]
Weight, kg	3.2 [1.9-3.8]
Maternal age, years	23 [16-38]
Weeks of gestation	37 [35-41]
Gender, n	
Male	25
Female	19
Primary diagnosis, n	
Aortic coarctation	39
Interrupted aortic arch	5
Associated heart defects, %	
Bicuspid aortic valve	27.3
Ventricular septal defect	47.7
Atrial septal defect	56.8

WOG = weeks of gestation.

Table 2: Echocardiographic data, intraoperative values, and surgical procedures.

Variable	Mean [range]	Percentiles P25, P50, P75
LVEF (%)	55 [20-80]	43, 56, 67
Z score		
Aortic valve	1.4 [−2.7-2.5]	−2.75, 1.8, 2.07
Transverse arch	1.8 [−2.7-2.5]	−2.2, 1.8, 2.0
Arch	−1.6 [−6-−2.3]	−2.4, −1.6, 1.95
Coarctation zone	−3 [−5.6-−1.5]	−3.9, −3.0, −2.3
Aortic cross-claming (min)	18 [15-30]	15, 17.5, 22
Total surgical time (min)	161 [50-290]	122.5, 165, 207
Variable	n (%)	
Aortic coartectomy	22 (50.0)	
Aortic advancement	13 (29.5)	
Graft placement	9 (9.1)	

LVEF = left ventricular ejection fraction.

The remainder were performed via lateral thoracotomy. No patients underwent correction of associated defects, only ligation of the ductus arteriosus.

Overall mortality was 13.6%, including 100% patients with aortic arch interruption. All patients who died were in the group admitted to the hospital more than 10 days before. No correlation was found between mortality and low weight. All patients who died had cardiogenic shock.

Regarding follow up, the average follows up time is 1.5 years, with a minimum of five months and a maximum of five years. To date, only two patients have presented with aortic recoarctation in the first six months after surgery, managed with interventional therapy. Both weighed less than 3 kg at the time of surgery.

DISCUSSION

Congenital heart defects are the most common birth abnormalities, with aortic arch obstructions being particularly prevalent in the neonatal period. Recognizing these defects is crucial as they fall under the category of critical heart diseases.[7] While specific clinical signs can aid detection, the initial presentation often involves shock, which can be mistaken for sepsis,[5] delaying diagnosis and management.

The primary associated cardiac conditions include patent ductus arteriosus, ventricular septal defect, and aortic stenosis. A bicuspid aortic valve is present in approximately two-thirds of patients, while mitral valve abnormalities are less common.[8] In our series, the most frequently associated pathology was an atrial septal defect, followed by a ventricular septal defect.

The management of aortic arch defects in neonates is primarily surgical. Some centers perform interventional management before surgery in patients with severe left ventricular dysfunction to improve function and reduce mortality. However, lacking a catheterization service at our center, all patients proceeded directly to surgery. Over 50% of our patient population presented with signs of cardiogenic shock, and all of them were referred to our facility when they were over 10 days old.

The definition of critical heart disease highlights that these are conduit-dependent heart diseases requiring timely management, ideally within the first 10 days of life, to reduce mortality.[5] Notably, transfer time was a significant variable in our study, with over 60% of cases transferred more than 10 days after birth. While the ideal timing for surgical correction of aortic coarctation in newborns depends on clinical manifestations, the descending aortic gradient, and ventricular function,[8] our findings indicate that late transfers significantly influence surgical timing, with an average of 15 days, thereby increasing patient morbidity and mortality.

Aortic arch defects are clinically suspected, but the initial diagnostic step is an echocardiogram, with other imaging techniques considered based on the findings. In our series, the mean gradient across the descending aorta was 54. The type of surgery performed is determined by aortic arch Z-values and anatomy. Our primary technique was simple anastomosis in 31.8% of cases (for simple aortic coarctation), while those with severe hypoplasia required graft placement (9.1%). All patients require follow-up, regardless of the surgical technique.

The increased risk of recoarctation in low birth weight patients has been widely discussed. A study by Bacha et al.[9] concluded that premature patients weighing less than 2 kg can undergo aortic coarctation repair with relatively low mortality. While the recoarctation rate remains significant in very low birth weight patients, it can be effectively treated with balloon dilation or reoperation. Notably, two patients in our series experienced recoarctation.

The overall mortality rate in our study was 13.6%, including 100% of patients with aortic arch interruption. All patients who died were admitted to the hospital after more than 10 days. No correlation was found between mortality and low weight. All deceased patients presented with cardiogenic shock, and it is important to mention that this level of care is not widely available in our state.

Despite our hospital being a reference center for the surgical management of these patients, the limited number of cases over five years is striking.

CONCLUSIONS

As previously mentioned, although we are a referral center in our state for the surgical care of patients with congenital heart disease, the number of cases in our series is limited;

furthermore, most patients were referred when they were older than ten days old, increasing their morbidity and mortality.

Despite significant technological advances in pediatric cardiology and cardiac surgery, a thorough physical examination remains essential for the early detection of congenital heart defects, ideally before birth. While not the primary focus of this study, it is worth noting that nationwide efforts are currently underway to implement mandatory cardiology screening. Although such screening is not without limitations, it undoubtedly represents a meaningful step forward compared to the absence of any early detection strategy, and plays a crucial role in improving the timely diagnosis and management of congenital heart disease.

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Funding: none.
Disclosure: the authors have no conflict of interest to disclose.