ORIGINAL ARTICLE

Aortic arch anomalies in pediatric patients: anatomic characteristics, associated malformations, and factors associated with surgical intervention at a national referral center in Peru, 2015–2024

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RESUMEN

Objective: To identify the types of aortic arch anomalies, describe associated congenital heart defects and extracardiac malformations, and evaluate factors associated with surgical intervention in pediatric patients treated at the Instituto Nacional de Salud del Niño San Borja during 2015–2024.
Methods: This was a retrospective observational study with an association analysis. Patients with aortic arch anomalies identified by cardiac computed tomography angiography were included. Demographic, anatomic, and clinical variables were recorded. Bivariate analyses and Firth's penalized multivariable logistic regression were performed, and crude odds ratios (ORs) and adjusted odds ratios (aORs) were estimated with their corresponding 95% confidence intervals (95% CIs).
Results: A total of 92 patients were included. The median age was 3 months (interquartile range [IQR]: 1–168 months). The left aortic arch was the most frequent configuration (52.2%), followed by the right aortic arch (44.6%). Hypoplasia was the most common obstructive abnormality (33.7%). Complex congenital heart disease was identified in 53.3% of patients and conotruncal congenital heart disease in 44.6%. Extracardiac malformations were observed in 52.2% of patients, with bronchopulmonary abnormalities being the most frequent (34.8%). Surgical intervention was recorded in 62.0% of cases. In the bivariate analysis, complex congenital heart disease, conotruncal congenital heart disease, and major obstructive aortic arch abnormalities were associated with surgical intervention. In the multivariable analysis, complex congenital heart disease (aOR = 3.98; 95% CI: 1.27–12.47; p = 0.018) and major obstructive aortic arch abnormalities (aOR = 8.16; 95% CI: 1.35–49.22; p = 0.022) remained independently associated with surgical intervention.
Conclusions: In this group of patients, the left aortic arch was the most frequent anatomic configuration, and hypoplasia was the most common obstructive abnormality of the aortic arch. Complex congenital heart disease and extracardiac malformations were frequent findings. Complex congenital heart disease and major obstructive aortic arch abnormalities were independently associated with surgical intervention.

Keywords: Aorta, Thoracic; Computed Tomography Angiography; Heart Defects, Congenital; Thoracic Surgery (Source: MeSH)

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Anomalías del arco aórtico en pacientes pediátricos: características anatómicas, malformaciones asociadas y factores asociados con la intervención quirúrgica en un centro de referencia nacional del Perú, 2015–2024

RESUMEN

Objetivo: Identificar los tipos de anomalías del arco aórtico, describir las cardiopatías congénitas y malformaciones extracardiacas asociadas, y evaluar los factores asociados con la realización de una intervención quirúrgica en pacientes pediátricos atendidos en el Instituto Nacional de Salud del Niño San Borja durante el periodo 2015–2024.

Métodos: Estudio observacional retrospectivo con análisis de asociación. Se incluyeron pacientes con anomalías del arco aórtico identificadas mediante angiografía por tomografía computarizada cardiaca. Se registraron variables demográficas, anatómicas y clínicas. Se realizaron análisis bivariados y una regresión logística multivariable penalizada de Firth, estimándose *odds ratios* crudos (OR) y ajustados (ORa) con sus respectivos intervalos de confianza al 95 % (IC 95 %).

Resultados: Se incluyeron 92 pacientes. La mediana de edad fue de 3 meses (RIC: 1–168 meses). Predominó el arco aórtico izquierdo (52,2 %), seguido del arco aórtico derecho (44,6 %). La hipoplasia fue la alteración obstructiva más común (33,7 %). Las cardiopatías congénitas complejas se identificaron en el 53,3 % de los pacientes y las conotruncales, en el 44,6 %. Se observaron malformaciones extracardiacas en el 52,2 % de los pacientes, siendo las más frecuentes las anomalías broncopulmonares (34,8 %). La intervención quirúrgica se registró en el 62,0 % de los casos. En el análisis bivariado, la cardiopatía congénita compleja, la cardiopatía conotruncal y las alteraciones obstructivas mayores del arco aórtico se asociaron con la realización de una intervención quirúrgica. En el análisis multivariable, la cardiopatía congénita compleja (ORa = 3,98; IC 95 %: 1,27–12,47; p = 0,018) y las alteraciones obstructivas mayores del arco aórtico (ORa = 8,16; IC 95 %: 1,35–49,22; p = 0,022) permanecieron asociadas de manera independiente con la realización de una intervención quirúrgica.

Conclusiones: En este grupo de pacientes, el arco aórtico izquierdo fue la configuración anatómica más frecuente y la hipoplasia fue la principal alteración obstructiva del arco aórtico. Las cardiopatías congénitas complejas y las malformaciones extracardiacas fueron hallazgos frecuentes. La cardiopatía congénita compleja y las alteraciones obstructivas mayores del arco aórtico se asociaron de manera independiente con la realización de una intervención quirúrgica.

Palabras clave: Aorta Torácica; Angiografía por Tomografía Computarizada; Cardiopatías Congénitas; Cirugía Torácica (Fuente: DeCS)

INTRODUCTION

Congenital aortic arch anomalies comprise a heterogeneous group of cardiovascular malformations resulting from disturbances in the embryonic development of the pharyngeal arches during the first weeks of gestation. These anomalies may occur as isolated anatomic variants or as part of congenital cardiovascular defects, including coarctation of the aorta, aortic arch hypoplasia, right aortic arch, double aortic arch, interrupted aortic arch, and aberrant subclavian artery, among others (1,2).

Congenital heart disease is the most common congenital anomaly in the pediatric population, with an estimated incidence of 8–10 per 1,000 live births (3). Within this group, aortic arch anomalies are relatively uncommon; however, their clinical significance lies in their association with other cardiovascular malformations and genetic syndromes. International studies have shown that certain anatomic variants, such as right aortic arch and interrupted aortic arch, frequently coexist with conotruncal defects, including tetralogy of Fallot, common arterial trunk, and ventricular septal defect (4,5).

Advances in cardiovascular imaging have substantially improved the diagnosis and anatomic characterization of these anomalies. Echocardiography remains the initial assessment tool in most pediatric patients; however, computed tomography angiography (CTA) and cardiovascular magnetic resonance provide detailed three-dimensional evaluation of vascular structures, enabling precise delineation of aortic arch anatomy, concomitant congenital heart disease, and associated extracardiac malformations (6,7). This information is essential for surgical planning and clinical follow-up of affected patients.

Several studies from Europe, North America, and Asia have described the anatomic distribution of aortic arch anomalies and their association with complex congenital heart disease (8–10). However, the available evidence from Latin America remains limited and heterogeneous (11). In Peru, reports of aortic arch anomalies in pediatric populations are scarce and consist mainly of case series or institutional reports, limiting knowledge of their distribution, predominant anatomic patterns, and associated malformations in this population (12,13).

The Instituto Nacional de Salud del Niño San Borja (INSN-SB) is a national referral center for the diagnosis and treatment of complex congenital heart disease and provides care to a large number of pediatric patients from different regions of the country. In this context, characterizing aortic arch anomalies, along with associated congenital heart disease and extracardiac malformations, may provide relevant epidemiologic data for future multicenter studies aimed at improving diagnostic algorithms and clinical stratification in this population. Likewise, identifying factors associated with surgical intervention could help identify subgroups of patients with greater need for specialized assessment and surgical planning.

Given the limited national evidence and the clinical importance of these anomalies in pediatric cardiovascular practice, there is a need to generate local data describing their anatomic characteristics, concomitant congenital heart disease, associated extracardiac malformations, and factors related to surgical intervention. Therefore, this study aimed to identify the types of aortic arch anomalies, describe associated congenital heart disease and extracardiac malformations, and evaluate factors associated with surgical intervention in pediatric patients treated at the INSN-SB during 2015–2024.

METHODS

Study design

A retrospective observational study with an association analysis was conducted based on the review of clinical information and cardiac computed tomography angiography (CTA) studies.

Population and sample

The study population consisted of patients younger than 18 years with aortic arch anomalies or variants diagnosed by CTA at the Diagnostic Imaging Service of the INSN-SB during 2015–2024. All patients who met the eligibility criteria during the study period were included.

Patients who had previously undergone cardiovascular surgery or intervention that substantially altered the original anatomy of the aortic arch before CTA were excluded. Patients whose studies had insufficient diagnostic image quality due to motion artifacts, inadequate vascular opacification, or incomplete anatomic coverage were also excluded. Finally, patients with insufficient data in their medical records were excluded. The process of participant identification, selection, exclusion, and inclusion is presented in Figure 1.

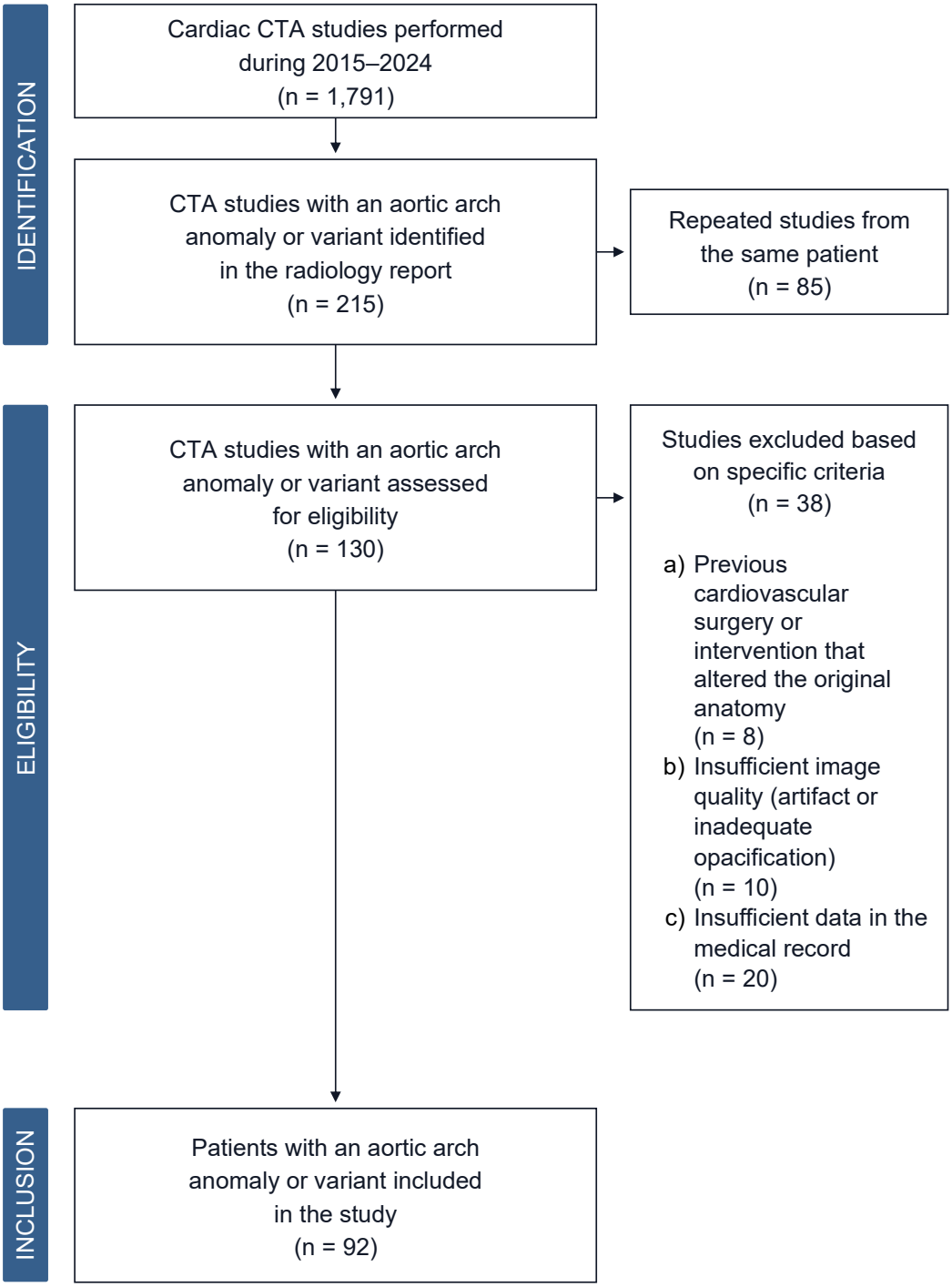

Figure 1. Flow diagram of the selection of patients included in the study

Procedures

Cases were identified by reviewing the radiology information system (RIS) database and the picture archiving and communication system (PACS). Patients whose CTA reports documented aortic arch anomalies or variants were selected. Duplicate studies from the same patient were removed, and the previously established inclusion and exclusion criteria were applied. For patients with more than one eligible study, only the first diagnostic study with adequate image quality was included.

All CTA studies were retrospectively reviewed by two radiologists with experience in cardiovascular imaging, who independently evaluated aortic arch anatomy and associated cardiovascular malformations. Aortic arch anomalies were classified using widely accepted anatomic criteria described in the specialized literature, including the identification of laterality variants, obstructive abnormalities, and associated vascular patterns. When discrepancies arose between observers regarding anatomic classification or the presence of associated anomalies, the images were jointly reevaluated until diagnostic consensus was reached.

Clinical and surgical information was obtained through retrospective review of electronic medical records and operative reports. All variables were recorded by one investigator using a data collection form specifically designed for the study. Data quality and consistency were assessed by identifying duplicate records and missing data and by verifying consistency among CTA findings, medical records, and operative reports.

For CTA evaluation, only studies with adequate multiplanar reconstructions, maximum-intensity projections, and three-dimensional volumetric reconstructions, as well as appropriate contrast-medium administration techniques and optimal acquisition parameters, were considered. Any inconsistencies identified were resolved by re-reviewing the original data sources before statistical analysis.

Variables

The independent variables included demographic characteristics, aortic arch laterality, supra-aortic vessel anomalies, obstructive aortic arch abnormalities, associated congenital heart disease, and extracardiac malformations. Age and sex were recorded as demographic variables. Age was recorded in months from birth to the time of evaluation.

Aortic arch laterality and supra-aortic vessel anomalies were evaluated by CTA, which was considered the reference standard for the anatomic characterization of these abnormalities. Aortic arch laterality was classified as left, right, or double aortic arch. Supra-aortic vessel anomalies were classified as isolated aberrant left subclavian artery (ALSA), aberrant right subclavian artery (ARSA), or ALSA associated with Kommerell diverticulum and a vascular ring. The anatomic classification was based on the patterns of persistence or regression of the pharyngeal arches described by Edwards and Van Praagh, which are widely used to explain the anatomic origin of these malformations (8).

Obstructive aortic arch abnormalities were classified as hypoplasia, coarctation, interruption of the aortic arch, other findings, or absence of an obstructive abnormality. For the bivariate and multivariable analyses, coarctation associated with aortic arch hypoplasia and interrupted aortic arch were defined as major obstructive aortic arch abnormalities.

Associated congenital heart disease was classified according to surgical complexity and embryologic criteria. According to surgical complexity, congenital heart disease was grouped into four categories: mild, moderate, complex, and no associated congenital heart disease. Mild congenital heart disease included atrial septal defect (ASD), small ventricular septal defect (VSD), and patent ductus arteriosus (PDA); moderate congenital heart disease included complex VSD and atrioventricular canal defects (AV canal); and complex congenital heart disease included tetralogy of Fallot (TOF), transposition of the great arteries (TGA), double-outlet right ventricle (DORV), common arterial trunk, pulmonary atresia with VSD, total anomalous pulmonary venous return (TAPVR), left ventricular hypoplasia, and single ventricle.

According to embryologic criteria, congenital heart disease was also classified as conotruncal, non-conotruncal, or absent. The conotruncal group included TOF, TGA, DORV, common arterial trunk, and pulmonary atresia with VSD. The non-conotruncal group included ASD, isolated VSD, AV canal, among others. Supplementary Table 1 describes the classification of all reported congenital heart defects according to embryologic criteria and surgical complexity.

Associated extracardiac malformations were grouped into six categories: bronchopulmonary, gastrointestinal, musculoskeletal, genetic, nephrourological, and neurological.

The dependent variable was cardiovascular surgical intervention after CTA, recorded dichotomously (yes/no) based on medical records and operative reports.

Statistical analysis

Statistical analyses were performed using IBM SPSS Statistics, version 25.0 (IBM Corp., Armonk, NY, USA). Categorical variables were reported as absolute frequencies and percentages. Quantitative variables were summarized using measures of central tendency and dispersion: mean and standard deviation for normally distributed variables, and median and interquartile range (IQR) when the assumption of normality was not met.

Bivariate analyses were performed to evaluate factors associated with surgical intervention. Anatomic and clinical variables were analyzed, including aortic arch laterality, obstructive aortic arch abnormalities, major obstructive abnormalities, the surgical complexity of congenital heart disease, and the embryologic classification of congenital heart disease. Associations between categorical variables were assessed using Pearson's chi-square test or Fisher's exact test. The magnitude of association was expressed as odds ratios (ORs) with 95% confidence intervals (CIs).

Subsequently, a multivariable logistic regression analysis was performed for explanatory purposes to identify variables independently associated with surgical intervention. Because of the low frequency of some anomalies and the risk of quasi-separation in the data, the multivariable analysis was performed using Firth's penalized logistic regression, a

technique that provides more stable estimates and reduces bias in small samples or when categories are infrequent. Independent variables were selected *a priori* based on clinical plausibility and previous evidence and included major obstructive aortic arch abnormalities, the presence of complex congenital heart disease, and conotruncal congenital heart disease. Results were expressed as adjusted odds ratios (aORs) with 95% CIs.

As exploratory sensitivity analyses, the potential confounding effect of age was evaluated using two additional conventional binary logistic regression models. In the first model, neonatal status was included as a dichotomous variable, with neonates defined as patients younger than 28 days at the time of CTA. In the second model, age was included as a continuous variable expressed in months. Both models retained the variables included in the main model: major obstructive aortic arch abnormalities, complex congenital heart disease, and conotruncal congenital heart disease. These analyses were performed to explore whether including age modified the associations observed in the main model.

A significance level of $p < 0.05$ was used for all tests.

Ethical considerations

This study respected patient anonymity and the confidentiality of information recorded in the medical records. Data were handled in coded form and used solely for research purposes. The study was conducted in accordance with the ethical principles of the World Medical Association Declaration of Helsinki, including the protection of life, health, dignity, integrity, the right to self-determination, and the privacy and confidentiality of participants' personal information. The study was approved by the Institutional Research Ethics Committee of the INSN-SB (code PI-957). The committee waived the requirement for informed consent because of the study's retrospective nature and minimal risk.

RESULTS

A total of 92 pediatric patients with aortic arch anomalies evaluated by CTA were included. The sex distribution was similar, with a slight male predominance (51.1%). The median age was 3 months (IQR: 1–168 months). CTA showed that the left aortic arch was the most frequent configuration (52.2%), followed by the right aortic arch (44.6%). The classic three-branch supra-aortic vessel configuration was present in 72.8% of patients, isolated ALSA in 12.0%, ARSA in 7.6%, and ALSA associated with Kommerell diverticulum and a vascular ring in 7.6%. Both presentations of ALSA were identified exclusively in patients with a right aortic arch, whereas ARSA was observed exclusively in patients with a left aortic arch. Regarding obstructive aortic arch abnormalities, hypoplasia was the predominant finding (33.7%), followed by combined coarctation and hypoplasia (12.0%), whereas interrupted aortic arch was observed in a smaller proportion of patients (4.3%).

Most patients had associated congenital heart disease, with complex congenital heart disease being the most frequent category (53.3%). From an embryologic perspective, conotruncal congenital heart disease accounted for 44.6% of cases. Extracardiac malformations were identified in 52.2% of patients. Bronchopulmonary abnormalities were the most frequent extracardiac malformations (32 patients; 34.8%), followed by gastrointestinal abnormalities (9 patients; 9.8%). A detailed description of aortic arch anatomy, associated congenital heart disease, and extracardiac malformations is provided in Table 1.

Table 1. Anatomic characteristics of the aortic arch, associated congenital heart disease, and extracardiac malformations in the study population (n = 92)

Finding	n (%)
Aortic arch laterality	
Left aortic arch	48 (52.2)
Right aortic arch	41 (44.6)
Double aortic arch	3 (3.3)
Supra-aortic vessels	
Normal	67 (72.8)
Isolated ALSA	11 (12.0)
ARSA	7 (7.6)
ALSA associated with Kommerell diverticulum and a vascular ring	7 (7.6)
Obstructive aortic arch abnormalities	
Normal	41 (44.6)
Hypoplasia	31 (33.7)
Coarctation associated with hypoplasia	11 (12.0)
Isolated coarctation	5 (5.4)
Interrupted aortic arch	4 (4.3)
Associated congenital heart disease according to surgical complexity	
Complex	49 (53.3)
Mild	29 (31.5)
Moderate	4 (4.3)
No associated congenital heart disease	10 (10.9)
Associated congenital heart disease according to embryologic criteria	
Conotruncal	41 (44.6)
Non-conotruncal	41 (44.6)
No associated congenital heart disease	10 (10.9)
Extracardiac malformations*	
Bronchopulmonary	32 (34.8)
Gastrointestinal	9 (9.8)
Genetic abnormalities	7 (7.6)
Musculoskeletal	7 (7.6)
Nephrourological	2 (2.2)
Neurological	1 (1.1)

*Patients could have extracardiac malformations in more than one category; therefore, the categories were not mutually exclusive. ALSA: aberrant left subclavian artery; ARSA: aberrant right subclavian artery.

Surgical intervention was recorded in 62.0% of patients. In the bivariate analysis, the presence of complex congenital heart disease was significantly associated with surgical intervention (p < 0.01) (Table 2), as was conotruncal congenital heart disease (p < 0.01) (Table 3). The overall analysis of obstructive aortic arch abnormalities across categories showed a statistically significant association with surgical intervention (p = 0.04) (Table 4). However, when the anatomically more complex lesions (coarctation associated with aortic arch hypoplasia and interrupted aortic arch) were grouped into a category termed "major obstructive abnormalities," a significant association with surgical intervention was observed (p = 0.02), although the wide confidence interval reflected the limited precision of the estimate due to the low frequency of these lesions (Table 5). No significant association was found between aortic arch laterality and surgical intervention (p = 0.359).

In the multivariable analysis using Firth's penalized logistic regression, major obstructive aortic arch abnormalities were associated with surgical intervention (aOR = 8.16; 95% CI: 1.35–49.22; p = 0.022). Complex congenital heart disease was also independently associated with surgical intervention (aOR = 3.98; 95% CI: 1.27–12.47; p = 0.018). The embryologic classification of congenital heart disease was not statistically significant after adjustment.

In the exploratory sensitivity analyses using conventional logistic regression, neonatal status was not associated with surgical intervention in the bivariate analysis (OR = 0.88; 95% CI: 0.36–2.16; p = 0.788) or after multivariable adjustment (aOR = 0.54; 95% CI: 0.18–1.63; p = 0.272) (Table 6). Similarly, age, entered as a continuous variable, was not independently associated with surgical intervention (aOR = 1.01 per 1-month increase; 95% CI: 0.99–1.03; p = 0.489) (Table 6). In both models, major obstructive aortic arch abnormalities and complex congenital heart disease maintained the direction and statistical significance of their associations, although the estimates were imprecise, as reflected by the width of the confidence intervals (Table 6).

Table 2. Bivariate analysis of complex congenital heart disease and surgical intervention

Complexity	Surgical intervention		OR (95% CI)	p
	No	Yes		
Complex	10	39	5.42 (2.20–13.60)	< 0.01
Non-complex	25	18		

Reference group: patients without complex congenital heart disease. OR: odds ratio; 95% CI: 95% confidence interval.

Table 3. Bivariate analysis of conotruncal congenital heart disease and surgical intervention

Classification	Surgical intervention		OR (95% CI)	p
	No	Yes		
Conotruncal	9	32	3.70 (1.47–9.29)	< 0.01
Non-conotruncal or no congenital heart disease	26	25		

Reference group: patients with non-conotruncal congenital heart disease or no congenital heart disease. OR: odds ratio; 95% CI: 95% confidence interval.

Table 4. Bivariate analysis of obstructive aortic arch abnormalities and surgical intervention

Abnormality	Surgical intervention		p
	No	Yes	
Normal	15	26	0.04
Hypoplasia	17	14	
Coarctation associated with hypoplasia	1	10	
Interrupted aortic arch	0	4	
Isolated coarctation	2	3	

The category "coarctation associated with hypoplasia" included patients who presented with both abnormalities. The p-value corresponds to the overall comparison among categories.

Table 5. Bivariate analysis of major obstructive aortic arch abnormalities and surgical intervention

Variable	Surgical intervention		OR (95% CI)	p
	No	Yes		
Major obstructive abnormalities	1	14	11.07 (1.40–88.40)	0.02
Other abnormalities	34	43		

The category "major obstructive abnormalities" included patients with coarctation associated with aortic arch hypoplasia or an interrupted aortic arch. Reference group: patients included in the "Other abnormalities" category. OR: odds ratio; 95% CI: 95% confidence interval.

Table 6. Multivariable analysis of factors associated with surgical intervention

Variable	aOR	95% CI	p
Major obstructive abnormalities	8.16	1.35–49.22	0.022
Complex congenital heart disease	3.98	1.27–12.47	0.018
Conotruncal congenital heart disease	1.69	0.53–5.42	0.377

Firth's penalized logistic regression was used because of the quasi-separation observed for the "major obstructive abnormalities" variable. aOR: adjusted odds ratio; 95% CI: 95% confidence interval.

DISCUSSION

In the present study, a high proportion of patients had associated congenital heart disease, particularly complex and conotruncal forms, and a considerable proportion underwent surgical intervention. These findings highlight the value of CTA in the comprehensive anatomic characterization of aortic arch anomalies and show that certain abnormalities, particularly major obstructive aortic arch abnormalities, were independently associated with surgical intervention.

The left aortic arch was the most frequent anatomic configuration, followed by the right aortic arch; a smaller proportion of patients had aberrant subclavian arteries and Kommerell diverticulum with a vascular ring. These results are consistent with those reported by Hanneman et al. (8), who described the left aortic arch as the predominant configuration even in populations with congenital heart disease, whereas right aortic arch variants and vascular rings represent a smaller but clinically relevant proportion because of their association with compressive symptoms and complex cardiovascular malformations. Similarly, Berdon et al. (14) noted that aortic arch anomalies constitute a heterogeneous spectrum of embryologic abnormalities whose clinical importance depends primarily on the associated structures and the presence of concomitant tracheoesophageal or cardiovascular involvement.

Likewise, the high frequency of complex congenital heart disease observed in this study is consistent with reports in the international literature. Several authors have shown that aortic arch anomalies are closely associated with conotruncal defects, particularly tetralogy of Fallot, common arterial trunk, double-outlet right ventricle, and transposition of the great arteries (5,15). McElhinney et al. (4) reported that the prevalence of aortic arch anomalies may exceed 20–30% in certain groups with conotruncal congenital heart disease, reinforcing the need for detailed anatomic evaluation before surgical intervention. The present study is consistent with these reports, as nearly half of the patients had congenital heart disease of conotruncal origin.

One of the most relevant findings of this study was the significant association between complex congenital heart disease and surgical intervention. This result was clinically expected, although its statistical demonstration provides local evidence of the prognostic importance of these malformations. Likewise, major obstructive aortic arch abnormalities, particularly combined coarctation and hypoplasia and interrupted aortic arch, showed the strongest independent association with surgical intervention. These results are consistent with those reported by Brown et al. (16) and Backer et al. (17), who described obstructive aortic arch lesions as one of the main indications for surgical reconstruction in pediatric patients because of their hemodynamic consequences and potential progression to heart failure or systemic arterial hypertension.

Conversely, although conotruncal congenital heart disease showed a significant association in the bivariate analysis, it did not remain statistically significant after adjustment. This finding may reflect that surgical intervention was primarily related to overall anatomic complexity and the presence of major obstructive aortic arch abnormalities rather than to the isolated embryologic classification of the congenital heart disease. This observation is consistent with Bae et al. (18), who emphasized that surgical planning depends on the complete cardiovascular anatomy rather than solely on the primary cardiac diagnosis.

Another relevant finding of the present study was the high frequency of associated extracardiac malformations, identified in more than half of the patients evaluated. Bronchopulmonary abnormalities were the predominant group, followed by gastrointestinal and genetic abnormalities. The bronchopulmonary abnormalities included left main bronchial stenosis, right upper lobe bronchial stenosis, tracheomalacia, pulmonary hypoplasia, and tracheal compression. However, these findings differ from those of other studies, such as Axt-Fliedner et al. (19), which reported a higher frequency of genetic abnormalities associated with aortic arch anomalies. This difference may be explained, at least in part, by the limited availability of comprehensive genetic testing in the population evaluated. In this regard, the coexistence of extracardiac malformations reinforces the concept that aortic arch anomalies should not be considered solely as isolated vascular abnormalities, but rather as part of a complex congenital phenotype.

From a diagnostic perspective, these results support the value of CTA as a reference method for the anatomic evaluation of the aortic arch. Although echocardiography is the initial study in most pediatric patients with suspected congenital heart disease, its limitations in evaluating extracardiac structures, supra-aortic vessels, and tracheobronchial relationships are well documented (19). In contrast, CTA provides high-spatial-resolution three-dimensional visualization, allows multiplanar reconstructions, and enables simultaneous assessment of vascular, cardiac, and mediastinal structures, all of which are essential for surgical planning (20,21).

The main clinical relevance of this study is that certain CTA findings may help identify patients who are more likely to require surgical intervention at the initial evaluation. In particular, identifying complex congenital heart disease and major obstructive aortic arch abnormalities could help guide multidisciplinary decision-making, procedure prioritization, and preoperative planning. Likewise, from a surgical perspective, the detailed anatomic characterization provided by CTA facilitates selection of the most appropriate surgical strategy and reduces intraoperative uncertainty.

This study represents one of the first national investigations to simultaneously evaluate aortic arch anatomy, associated cardiovascular malformations, and factors associated with surgical intervention in a pediatric population treated at a national referral center. The incorporation of multivariable analysis provides a more robust approach than that used in many previous descriptive studies and allows the identification of clinically relevant variables for patient care.

One of the study's strengths was the use of CTA records to simultaneously evaluate cardiac and extracardiac structures, allowing the identification of relevant structural abnormalities that might not be fully characterized by other diagnostic modalities. Other strengths included the inclusion of all eligible patients treated during the study period at a national high-complexity center and the analysis of anatomic and clinical variables with potential prognostic value.

However, several limitations should be considered. First, this was a retrospective single-center study, which may limit the generalizability of the results. Second, the relatively small sample size reduces the statistical power to detect associations in some infrequent anatomic subcategories, such as double aortic arch or interrupted aortic arch. Third, a conceptual limitation of the present study is the possibility of incorporation bias, because major obstructive aortic arch

abnormalities identified by CTA often constitute the primary anatomic basis for surgical intervention. In this context, the observed association between these variables and surgical intervention should not be interpreted as an independent prognostic model, but rather as a description of the anatomic findings underlying the indication for surgical treatment in clinical practice. Moreover, the observational nature of the study precluded establishing causal relationships between the variables analyzed. Fourth, another limitation arose from the case identification method. Patients were selected based on radiology reports documenting aortic arch anomalies; therefore, it cannot be ruled out that some studies with anatomic abnormalities that were not initially reported were excluded. This may have introduced detection bias and led to underestimation of the true frequency of these anomalies. Nevertheless, because this was a national referral center for congenital heart disease, where studies were interpreted by radiologists experienced in pediatric cardiovascular imaging, this bias was likely limited.

CONCLUSION

In this series of pediatric patients, the left aortic arch was the most frequent anatomic configuration, and hypoplasia was the most common obstructive abnormality. Complex congenital heart disease and extracardiac malformations, particularly bronchopulmonary abnormalities, were frequent findings. Complex congenital heart disease and major obstructive aortic arch abnormalities were independently associated with surgical intervention. CTA provided detailed anatomic characterization of the aortic arch and associated malformations, providing relevant information for surgical planning and multidisciplinary management.

Author contributions

BBZ: Conceptualization, Methodology, Writing – review and editing. GBU: Formal analysis, Writing – original draft. All authors approved the final version of the manuscript.

Conflicts of interest

The authors declare no relevant financial or non-financial conflicts of interest.

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Data availability

The data supporting the findings of this study are available upon request from the corresponding author.

REFERENCES

1. Backer CL, Rigsby CK, Mavroudis C. Vascular rings and pulmonary artery sling. In: Mavroudis C, Backer CL, editors. Pediatric cardiac surgery. 5th ed. Chichester (UK): Wiley-Blackwell; 2023. p. 225-47. doi: 10.1002/9781119282327.ch12
2. Asir M, Brennan K, Heraghty J, Hurn A, Jogeesvaran H, Moriarty G, et al. Vascular rings in the current era: is this a new disease requiring a new approach? Arch Dis Child Educ Pract Ed. 2026;111(1):23-31. doi: 10.1136/archdischild-2024-327517
3. van der Linde D, Konings EEM, Slager MA, Witsenburg M, Helbing WA, Takkenberg JJM, et al. Birth prevalence of congenital heart disease worldwide: a systematic review and meta-analysis. J Am Coll Cardiol. 2011;58(21):2241-7. doi: 10.1016/j.jacc.2011.08.025
4. McElhinney DB, Clark BJ 3rd, Weinberg PM, Kenton ML, McDonald-McGinn D, Driscoll DA, et al. Association of chromosome 22q11 deletion with isolated anomalies of aortic arch laterality and branching. J Am Coll Cardiol. 2001;37(8):2114-9. doi: 10.1016/S0735-1097(01)01286-4
5. Weinberg PM. Aortic arch anomalies. J Cardiovasc Magn Reson. 2006;8(4):633-43. doi: 10.1080/10976640600713756
6. Epstein R, Yomogida M, Donovan D, Butensky A, Aidala AA, Farooqi KM, et al. Trends in cardiac CT utilization for patients with pediatric and congenital heart disease: a multicenter survey study. J Cardiovasc Comput Tomogr. 2024;18(3):267-73. doi: 10.1016/j.jcct.2024.02.002
7. Francone M, Gimelli A, Budde RPJ, Caro-Dominguez P, Einstein AJ, Gutberlet M, et al. Radiation safety for cardiovascular computed tomography imaging in paediatric cardiology: a joint expert consensus document of the EACVI, ESCR, AEPC, and ESPR. Eur Heart J Cardiovasc Imaging. 2022;23(8):e279-89. doi: 10.1093/ehjci/jeac048
8. Hanneman K, Newman B, Chan F. Congenital variants and anomalies of the aortic arch. Radiographics. 2017;37(1):32-51. doi: 10.1148/rg.2017160033
9. Priya S, Thomas R, Nagpal P, Sharma A, Steigner M. Congenital anomalies of the aortic arch. Cardiovasc Diagn Ther. 2018;8(Suppl 1):S26-44. doi: 10.21037/cdt.2017.10.15
10. Razon Y, Berant M, Fogelman R, Amir G, Birk E. Prenatal diagnosis and outcome of right aortic arch without significant intracardiac anomaly. J Am Soc Echocardiogr. 2014;27(12):1352-8. doi: 10.1016/j.echo.2014.08.003
11. Ibáñez-Correa LM, Victoria S, Hurtado-Villa P. Prevalencia de cardiopatías congénitas en una cohorte de 54.193 nacimientos entre 2011-2017. Rev Colomb Cardiol. 2021;28(1):53-9. doi: 10.24875/RCCAR.M21000009
12. Lacunza Paredes RO, Sembrera Palacios E. Serie de casos de arco aórtico derecho: aproximación diagnóstica prenatal. Rev Peru Ginecol Obstet. 2022;68(1). doi: 10.31403/rpgo.v68i2403
13. Davila-Flores D, Villasante-Villalta C, Montesinos-Segura R, Zúñiga-Meza J, Peralta-Santos H, Vera-Talledo L. Anillo vascular tipo doble arco aórtico como causa de estridor y disfagia en la infancia: reporte de caso. Arch Peru Cardiol Cir Cardiovasc. 2025;6(4):253-7. doi: 10.47487/apcyccv.v6i4.534
14. Berdon WE, Baker DH, Wung JT, Chrispin A, Kozlowski K, de Silva M, et al. Complete cartilage-ring tracheal stenosis associated with anomalous left pulmonary artery: the ring-sling complex. Radiology. 1984;152(1):57-64. doi: 10.1148/radiology.152.1.6729137
15. Ho SY, Wilcox BR, Anderson RH, Lincoln JCR. Interrupted aortic arch--anatomical features of surgical significance. Thorac Cardiovasc Surg. 1983;31(4):199-205. doi: 10.1055/s-2007-1021980

16. Brown JW, Ruzmetov M, Okada Y, Vijay P, Rodefeld MD, Turrentine MW. Outcomes in patients with interrupted aortic arch and associated anomalies: a 20-year experience. Eur J Cardiothorac Surg. 2006;29(5):666-73. doi: 10.1016/j.ejcts.2006.01.060
17. Backer CL, Mongé MC, Popescu AR, Eltayeb OM, Rastatter JC, Rigsby CK. Vascular rings. Semin Pediatr Surg. 2016;25(3):165-75. doi: 10.1053/j.sempedsurg.2016.02.009
18. Bae SB, Kang EJ, Choo KS, Lee J, Kim SH, Lim KJ, et al. Aortic arch variants and anomalies: embryology, imaging findings, and clinical considerations. J Cardiovasc Imaging. 2022;30(4):231-62. doi: 10.4250/jcvi.2022.0058
19. Axt-Fliedner R, Nazar A, Bedei I, Schenk J, Reitz M, Rupp S, et al. Associated anomalies and outcome in patients with prenatal diagnosis of aortic arch anomalies as aberrant right subclavian artery, right aortic arch and double aortic arch. Diagnostics (Basel). 2024;14(3):238. doi: 10.3390/diagnostics14030238
20. Cohen J, Asrani P, Lee S, Frush D, Han BK, Chelliah A, et al. Cardiovascular computed tomography in pediatric congenital heart disease: a state of the art review. J Cardiovasc Comput Tomogr. 2022;16(6):467-82. doi: 10.1016/j.jcct.2022.04.004
21. Dillman JR, Attili AK, Agarwal PP, Dorfman AL, Hernandez RJ, Strouse PJ. Common and uncommon vascular rings and slings: a multimodality review. Pediatr Radiol. 2011;41(11):1440-54; quiz 1489-90. doi: 10.1007/s00247-011-2131-2