ORIGINAL RESEARCH

Coarctation of the Aorta and Bicuspid Aortic Valve in Pediatric Patients – a Single-center Study and Literature Review

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ABSTRACT

Background: This study focuses on two of the most prevalent congenital heart defects: bicuspid aortic valve (BAV) and coarctation of the aorta (CoA). In severe forms, CoA is an emergency that can become life-threatening. Both diseases are linked to aortic vasculopathy, which represents a significant contributor to morbidity and mortality in young patients with congenital heart disease. The aim of this study was to investigate whether there are differences between patients with CoA and BAV, and patients with CoA and without BAV in terms of outcome and echocardiographic parameters, and to evaluate the efficacy of different echocardiographic parameters in predicting CoA. **Methods:** A retrospective and prospective observational analytic study was conducted between January 2018 and October 2024 at a tertiary pediatric cardiology referral center enrolling patients aged between 0 and 17 years, diagnosed with CoA, who underwent surgical CoA repair. The common carotid-subclavian artery index (CSAI) and the aortic isthmus-descending aorta index (I/D) were calculated based on echocardiographic measurements. **Results:** A total of 101 patients underwent surgical or interventional treatment for CoA in the studied period, 58.4% being neonates. Based on echocardiographic examination, 68.3% of patients had BAV with CoA. The mean value of the carotid-subclavian index was 0.95 in neonates, compared to 1.11 in children (p = 0.035). No statistically significant difference was observed in the mean value of the aortic isthmus-descending aorta index between the two groups (p = 0.237). **Conclusions:** The implications of the coexistence of the two conditions are not yet fully understood. This study did not find significant echocardiographic differences between patients with CoA and BAV and patients with CoA and without BAV. Further long-term, multicenter, multi-arm studies with larger groups are required in these areas.

Keywords: coarctation of the aorta, bicuspid aortic valve, CSAI

INTRODUCTION

The prevalence of congenital heart disease (CHD) has exhibited a consistent upward trajectory over the past few decades. Coarctation of the aorta (CoA) and bicuspid aortic valve (BAV) are associated with aortic vasculopathy, a significant contributor to morbidity and mortality in young patients with CHD.

BAV is one of the most common forms of CHD, with higher prevalence in men. The most frequently observed phenotype of BAV in conjunction with CoA is type IA (fusion of the left and right coronary cusps).[1,2]

CoA is a congenital narrowing of the aortic isthmus, frequently observed as an isolated stenosis or as a longer hypoplastic segment of the ascending aorta. The prevalence of CoA in relation to all CHDs is 4–8%, while the isolated form of CoA has a prevalence of 3 in 10,000 live births.[1,3]

It is frequently the case that both BAV and CoA as CHD are present concurrently, occurring in 85% of cases.[4] In cases of CoA, the establishment of a therapeutic approach is the responsibility of a multidisciplinary team. A variety of surgical techniques have been used for the repair of CoA. The mortality and morbidity rates associated with surgical intervention are relatively low. For instance, a recent study reported a 30-day mortality rate of only 0.54%. It is acknowledged that complications may arise, including recoarctation at either an early or late stage, persistent hypertension, and heart failure.[5] Although surgical intervention remains the primary treatment for children diagnosed with CoA, percutaneous intervention may be used as a strategy for high-risk neonates as a definitive treatment or as a bridge to surgery. The optimal management strategy for CoA remains a topic of debate, and there is currently no comprehensive evidence-based standard of care or algorithm.[6,7]

To accurately assess the risks associated with patient populations with CoA, it is important to consider not only the severity of CoA, but also the prevalence and severity of mixed valvular disease.[8,9] Aortopathy may be a significant contributor to the increased morbidity and mortality observed in both conditions.[10,11]

Objective

This study was designed to investigate whether there are differences between patients with CoA and BAV, and patients with CoA and without BAV in terms of outcome and echocardiographic parameters. The second aim of this study was to evaluate the efficacy of different echocardiographic parameters in predicting CoA.

MATERIAL AND METHOD

A retrospective and prospective observational analytical study was conducted at a tertiary pediatric cardiology referral center between January 2018 and October 2024, enrolling patients aged 0–17 years diagnosed with CoA who underwent surgical CoA repair. The patients were divided into two primary categories: Group A, consisting of individuals with CoA and BAV, and Group B, consisting of individuals with CoA but without BAV. Demographic parameters including age, weight, height, body surface area, associated anomalies or intracardiac surgery at the time of CoA surgery, and surgical procedure were meticulously documented. All echocardiograms were analyzed by a single experienced pediatric echocardiographer who was blinded to the study outcome at the time of image analysis.

Exclusion criteria were: lack of follow-up data and/or lack of available predischarge echocardiograms of adequate quality, genetic syndromes, metabolic disorders, oncological diagnoses, liver disease, chronic respiratory or renal disease.

Cardiac ultrasound evaluation

The acquired images were stored in DICOM format and then subjected to offline analysis. The aortic valve was evaluated by analyzing the parasternal short-axis view. The parasternal long-axis view was used to calculate Z-scores perpendicular to the long axis of the aorta using the inner edge technique.[12,13] The suprasternal view was used to evaluate the diameter of the innominate artery, proximal aortic arch, left common carotid artery, transverse aortic arch, aortic isthmus, and of the descending aorta at the level of the diaphragm; the distance between the innominate artery and the left common carotid artery, between the left common carotid artery and the left subclavicular artery; and persistence of diastolic gradient in the descending aorta. The presence or absence of a patent ductus arteriosus was determined and, if present, the diameter of the arterial duct and the direction of the shunt through the patent ductus arteriosus were described.

Based on the echocardiographic measurements, both the common carotid-subclavian artery index (CSAI) and the aortic isthmus-descending aorta index (I/D) were calculated according to the previously described methodology. CSAI is the ratio of the aortic arch diameter at the level of the subclavian artery to the distance between the left common carotid artery and the left subclavian artery. Meanwhile, I/D is the ratio between the diameters of the aortic isthmus and the descending aorta.[14,15]

TABLE 1. The characteristics of patients with CoA

Variable	n = 101
Age (months, median, IQR_{25}-IQR_{75})	0.00 (0.00–5.00)
Newborn (n, %)	59 (58.4%)
Weight (kg, median, IQR_{25}-IQR_{75})	3.99 (3.24–6.22)
Length (cm, median, IQR_{25}-IQR_{75})	56.00 (52.00–65.00)
BSA (m², median, IQR_{25}-IQR_{75})	0.24 (0.21–0.32)
Male (n, %)	65 (64.35%)
Female (n, %)	36 (35.64%)
Antenatal diagnosis (n, %)	41 (40.6%)
Systolic blood pressure preductal (mmHg, mean ± s.d.)	102.26 ± 21.79
Systolic blood pressure postductal (mmHg, mean ± s.d.)	85.38 ± 17.92

TABLE 2. Associated congenital heart lesions

Congenital heart lesions	n (%)
BAV	69 (68.3%)
Atrial septal defect/Foramen ovale patency	77 (76.2%)
Ventricular septal defect	31 (30.7%)
Patent ductus arteriosus	60 (59.4%)
Subvalvular aortic stenosis	10 (9.9%)
Valvular aortic stenosis	12 (11.9%)
Double outlet of right ventricle	1 (1.0%)
Aortic arch hypoplasia	44 (43.6%)
Borderline left ventricle	12 (11.9%)
Left superior vena cava	8 (7.9%)

Statistical analysis

The statistical analysis was conducted using the R statistical computing platform, and graphical representations were generated with the matplotlib Python library for data visualization.[16,17] Categorical data were presented as numbers and percentages, whereas continuous parametric data were expressed as mean ± s.d., and non-parametric continuous data as median (interquartile range). The assumption of normality was evaluated through the Kolmogorov–Smirnov test. Comparisons of central tendency were conducted using t-tests for parametric data and Mann–Whitney tests for non-parametric data. In instances where multiple groups were involved, a one-way analysis of variance (ANOVA) was used for parametric data, with post-hoc Tukey tests subsequently conducted. For non-parametric data, the Kruskal–Wallis test was used, with Dunn's test for post-hoc analysis. Frequency comparisons were performed using variations of the chi-squared test. A significance level of α = 0.05 was considered for all analyses.

Ethics

The research was conducted in accordance with the principles set forth in the Declaration of Helsinki. The research protocol was approved by the Ethics Committee of "George Emil Palade" University of Medicine, Pharmacy, Science and Technology of Târgu Mureș (approval no. 3359/07.10.2024).

RESULTS

A total of 101 patients underwent surgical or interventional treatment for CoA between January 2018 and October 2024. Of the aforementioned patients, 58.4% were neonates. The characteristics of these patients are shown in Table 1. Based on the echocardiographic examination, 68.3% of patients had BAV with CoA (Table 2). Diastolic flow pattern was found in 79.2% of patients, with a pressure gradient in the coarctation region with a median of 36 mmHg (Table 3).

The primary objective of the study was to investigate whether there are differences between patients with CoA and BAV, and patients with CoA and without BAV. The results demonstrated no statistically significant differences between the two groups, with the exception of the Z-score of the distal ascending aorta (p = 0.013; Table 4). In relation to the second objective of this study, which was to evaluate and compare the predictive efficacy of different echocardiographic parameters in identifying cases of CoA, a comparison was made between the CSAI and the I/D index. The results of this analysis indicated no statistically significant differences between the two groups (Table 4).

In view of the inhomogeneity of the group and the high percentage of newborns, an additional data set was obtained through an analysis of the demographic data, associated lesions, and the echocardiographic parameters described above. The mean value of the CSAI was 0.95 in neonates, compared to 1.11 in children (p = 0.035). No statistically significant difference was observed in the mean value of the I/D index between the two groups (p = 0.237; Tables 5, 6, and 7).

TABLE 3. Echocardiographic findings of the patients

Variable	Value
Diastolic flow pattern (n, %)	80 (79.2)
Pressure gradient (median, IQR_{25}-IQR_{75})	36.00 (18.00 – 65.25)
Ejection fraction (mean ± s.d., %)	68.47 ± 9.88

TABLE 4. Comparison between absolute size and *Z*-score of echocardiographic parameters of the two groups

Variable	Group A (CoA + BAV)	Group B (CoA)	p value
Aortic valve annulus (mm, median, IQR_{25}-IQR_{75})	6.50 (6.00–8.40)	7.19 (5.90–9.90)	0.411
Aortic valve annulus *Z*-score (mean ± s.d.)	−0.96 ± 2.06	−0.48 ± 1.74	0.243
Sinus of Valsalva (mm, median, IQR_{25}-IQR_{75})	8.79 (7.55–11.15)	8.80 (7.40–11.30)	0.822
Sinus of Valsalva *Z*-score (mean ± s.d.)	−1.25 ± 2.12	−1.49 ± 1.92	0.612
Sino-tubular junction (mm, median, IQR_{25}-IQR_{75})	7.33 (6.47–8.98)	8.52 (6.67–9.97)	0.134
Sino-tubular junction *Z*-score (mean ± s.d.)	−0.63 ± 1.93	−0.15 ± 1.51	0.23
Ascending aorta (mm, median, IQR_{25}-IQR_{75})	8.00 (6.90–10.40)	9.34 (7.50–10.65)	0.273
Ascending aorta *Z*-score (mean ± s.d.)	−1.53 (−2.99–0.34)	−0.88 (−1.63–0.71)	0.119
Right innominate artery diameter (mm, median, IQR_{25}-IQR_{75})	4.45 (4.00–5.83)	5.10 (4.45–6.09)	0.285
Distal ascending aorta diameter (mm, median, IQR_{25}-IQR_{75})	6.43 (5.15–8.85)	8.00 (6.45–10.28)	0.068
Distal ascending aorta diameter *Z*-score (mean ± s.d.)	−1.20 ± 2.40	0.23 ± 2.24	0.013
Proximal transverse arch diameter (mm, median, IQR_{25}-IQR_{75})	5.35 (3.90–7.60)	6.15 (5.26–7.70)	0.123
Distance between the right innominate artery and left common carotid artery (mm, median, IQR_{25}-IQR_{75})	3.31 (2.29–4.97)	4.31 (3.62–5.71)	0.033
Left common carotid artery diameter (mm, median, IQR_{25}-IQR_{75})	3.80 (3.10–4.79)	4.25 (3.19–5.60)	0.456
Distal transverse arch (mm, median, IQR_{25}-IQR_{75})	5.00 (3.80–6.10)	5.20 (4.10–9.80)	0.13
Distal transverse arch *Z*-score (mean ± s.d.)	−2.63 (−4.36 to −1.41)	−2.45 (−3.80 to −1.81)	0.97
Distance between the left common carotid artery and the left subclavian artery (mm, median, IQR_{25}-IQR_{75})	5.12 (3.85–7.00)	6.30 (3.70–7.32)	0.55
CSAI	0.96 (0.68–1.29)	1.02 (0.72–1.63)	0.49
Aortic isthmus (mm, median, IQR_{25}-IQR_{75})	2.60 (2.10–3.20)	3.00 (2.40–4.00)	0.068
Aortic isthmus *Z*-score (mean ± s.d.)	−5.39 ± 1.82	−4.83 ± 1.94	0.19
Descending aorta at the diaphragm (mm, median, IQR_{25}-IQR_{75})	5.45 (4.33–6.67)	6.10 (5.30–7.90)	0.125
I/D index	0.45 (0.36–0.53)	0.53 (0.42–0.60)	0.179
Patent ductus arteriosus (mean ± s.d.)	3.73 ± 1.50	2.14 ± 0.96	<0.001

The study participants were divided into subgroups based on their CSAI and I/D index values. Patients with a CSAI value of less than 1.5 exhibited diminished absolute size and *Z*-score values of the ascending aorta, proximal aortic arch, and aortic isthmus. Additionally, subjects with CSAI values below 1.5 exhibited a greater distance between the common carotid artery and the subclavian artery. It is noteworthy that the absolute value of the pulmonary artery trunk was also observed to be lower. Of the subjects with a CSAI <1.5, 63.9% were neonates, 73.8% had BAV, and 18% had aortic recoarctation (Table 8).

In terms of the I/D index, statistically significant data were obtained for absolute size and *Z*-score from the ascending aorta, aortic isthmus, and descending aorta at the level of the diaphragm. Among the subjects with values less than 0.64, 59% were neonates, 70.5% had BAV, and 19.7% had aortic recoarctation (Table 9).

With regard to surgical correction in both newborns and children, the most common procedure was resection and terminal–terminal anastomosis. The next most common surgical intervention was widening of the aorta using a heterologous pericardial patch or Cormatrix.

TABLE 5. The characteristics of patients with CoA

Variable	Group A (CoA + BAV)	Group B (CoA)
Age (months, median, IQR_{25}-IQR_{75})	0.00 (0.00–0.00)	8.00 (2.00–70.00)
Weight (kg, median, IQR_{25}-IQR_{75})	3.50 (3.00–3.98)	7.20 (4.50–19.60)
Length (cm, median, IQR_{25}-IQR_{75})	54.00 (52.00–56.25)	69.00 (58.50–118.50)
BSA (m^2, median, IQR_{25}-IQR_{75})	0.22 (0.20–0.24)	0.36 (0.27–0.81)
Antenatal diagnosis (n, %)	33 (55.9%)	8 (19.0%)
Systolic blood pressure preductal (mmHg, mean ± s.d.)	93.02 ± 17.24	114.14 ± 21.42
Systolic blood pressure postductal (mmHg, mean ± s.d.)	79.43 ± 15.43	93.42 ± 18.08

TABLE 6. Frequency of associated CHD

Congenital heart lesions	Newborn (*n* = 59)	Child (*n* = 42)	p value
BAV	43 (72.9%)	26 (61.9%)	0.341
Patent ductus arteriosus	47 (79.7%)	13 (31.0%)	<0.001
Atrial septal defect/Foramen ovale patency	55 (93.2%)	22 (52.4%)	<0.001
Ventricular septal defect	21 (35.6%)	10 (23.8%)	0.295
Aortic arch hypoplasia	34 (57.6%)	10 (23.8%)	0.002
Borderline left ventricle	10 (16.9%)	2 (4.8%)	0.116
Left superior vena cava	6 (10.2%)	2 (4.8%)	0.464

Thirteen patients with an associated ventricular septal defect required surgical repair of the coarctation and pulmonary artery banding. Four patients underwent aortoplasty with a left subclavian artery flap. In terms of interventional treatment, a total of 12 patients underwent this procedure, including two patients who received balloon angioplasty and 10 who received stent implantation. The most frequently observed postoperative complications were aortic recoarctation (20.0%) and secondary arterial hypertension (62.4%). In the entire cohort of 59 newborns, 13 presented with aortic recoarctation (22%), with no statistically significant differences between the newborn and child group (p = 0.72). In the cohort of patients who underwent surgical correction, 6 patients exhibited recoarctation following the terminal–terminal procedure. Four patients displayed recoarctation after surgery involving heterologous pericardial patch widening. Additionally, two patients experienced recoarctation after undergoing stent implantation, and one patient following balloon angioplasty. In total, 32 of the newborns exhibited secondary hypertension, and 31 of the children in the control group also displayed this phenomenon; the difference was not statistically significant.

TABLE 7. Echocardiographic parameters

Congenital heart lesions	Newborn (*n* = 59)	Child (*n* = 42)
Aortic valve annulus (mm, median, IQR_{25}-IQR_{75})	6.00 (5.50–6.50)	9.90 (7.80–15.50)
Aortic valve annulus *Z*-score (mean ± s.d.)	−1.59 ± 1.51	0.30 ± 2.06
Sinus of Valsalva (mm, median, IQR_{25}-IQR_{75})	8.20 (6.90–8.90)	12.50 (9.30–19.95)
Sinus of Valsalva *Z*-score (mean ± s.d.)	−2.03 ± 1.76	−0.19 ± 2.00
Sino-tubular junction (mm, median, IQR_{25}-IQR_{75})	6.70 (6.26–7.92)	10.00 (8.09–16.90)
Sino-tubular junction *Z*-score (mean ± s.d.)	−0.94 ± 1.63	0.21 ± 1.91
Ascending aorta (mm, median, IQR_{25}-IQR_{75})	7.60 (6.90–9.00)	11.60 (8.49–20.00)
Ascending aorta *Z*-score (mean ± s.d.)	−1.47 ± 1.91	−0.05 ± 2.68
Right innominate artery (mm, median, IQR_{25}-IQR_{75})	4.56 ± 1.03	6.16 ± 2.68
Distal ascending aorta (mm, median, IQR_{25}-IQR_{75})	6.29 ± 1.97	11.23 ± 5.63
Distal ascending aorta *Z*-score (mean ± s.d.)	−1.28 ± 2.56	−0.04 ± 2.04
Proximal transverse arch (mm, median, IQR_{25}-IQR_{75})	5.10 (3.90–6.70)	7.30 (5.65–13.80)
Distance between the right innominate and left common carotid artery diameter (mm, median, IQR_{25}-IQR_{75})	3.10 (2.25–4.40)	5.20 (4.00–6.34)
Left common carotid artery diameter (mm, median, IQR_{25}-IQR_{75})	3.37 (3.00–4.00)	4.70 (3.92–6.52)
Distal transverse arch (mm, median, IQR_{25}-IQR_{75})	4.40 ± 1.11	7.80 ± 3.47
Distal transverse arch *Z*-score (mean ± s.d.)	−2.99 (−4.51 to −1.96)	−1.88 (−3.31 to −1.06)
Distance between the left common carotid artery and the left subclavian artery (mm, median, IQR_{25}-IQR_{75})	4.90 (3.59–6.61)	6.40 (4.70–7.75)
CSAI (median, IQR_{25}-IQR_{75})	0.95 (0.61–1.29)	1.11 (0.90–1.58)
Aortic isthmus (mm, median, IQR_{25}-IQR_{75})	2.60 (2.20–3.00)	3.20 (2.20–4.00)
Aortic isthmus *Z*-score (mean ± s.d.)	−4.78 ± 1.90	−5.92 ± 1.59
Descending aorta at diaphragm (mm, median, IQR_{25}-IQR_{75})	5.35 (4.29–6.17)	7.10 (5.04–9.77)
I/D index (median, IQR_{25}-IQR_{75})	0.48 (0.38–0.56)	0.42 (0.29–0.56)

TABLE 8. Comparative subgroup analysis according to the CSAI

Variable	Low (≤ 1.5)	High (> 1.5)	p value
Age (months, median, IQR$_{25}$-IQR$_{75}$)	0.00 (0.00–2.00)	2.00 (0.00–36.00)	0.07
Weight (kg, median, IQR$_{25}$-IQR$_{75}$)	3.86 (3.19–5.03)	4.40 (3.40–12.50)	0.226
Length (cm, median, IQR$_{25}$-IQR$_{75}$)	55.00 (52.00–62.00)	59.00 (52.00–94.00)	0.372
BSA (m2, median, IQR$_{25}$-IQR$_{75}$)	0.23 (0.21–0.29)	0.25 (0.21–0.57)	0.308
Distal ascending aorta diameter (mm, median, IQR$_{25}$-IQR$_{75}$)	7.08 (5.51–8.90)	8.80 (6.80–12.30)	0.113
Distal ascending aorta Z-score (median, IQR$_{25}$-IQR$_{75}$)	−1.00 (−2.80–0.56)	0.61 (0.01–1.32)	0.02
Proximal transverse arch (mm, median, IQR$_{25}$-IQR$_{75}$)	5.91 (3.93–7.30)	7.00 (5.20–12.40)	0.027
Distance between the right innominate and left common carotid artery (mm, median, IQR$_{25}$-IQR$_{75}$)	3.88 (2.56–5.06)	4.89 (3.00–6.34)	0.344
Left common carotid artery (mm, median, IQR$_{25}$-IQR$_{75}$)	3.80 (3.10–4.54)	4.40 (3.60–5.96)	0.146
Distal transverse arch (mm, median, IQR$_{25}$-IQR$_{75}$)	4.60 (3.64–5.31)	7.30 (5.20–10.30)	<0.001
Distal transverse arch Z-score (median, IQR$_{25}$-IQR$_{75}$)	−2.79 (−4.50 to −1.52)	−1.73 (−2.80 to −0.47)	0.018
Distance between left common carotid artery and the left subclavian artery (mm, median, IQR$_{25}$-IQR$_{75}$)	6.20 (4.40–7.58)	3.60 (3.00–5.00)	<0.001
CSAI (median, IQR$_{25}$-IQR$_{75}$)	0.86 (0.62–1.06)	1.80 (1.63–2.24)	<0.001
Aortic isthmus (mm, median, IQR$_{25}$-IQR$_{75}$)	2.60 (2.20–3.15)	3.40 (2.50–3.92)	0.025
Aortic isthmus Z-score (median, IQR$_{25}$-IQR$_{75}$)	−5.18 (−6.32 to −3.74)	−4.56 (−5.07 to −4.13)	0.412
I/D (median, IQR$_{25}$-IQR$_{75}$)	0.47 (0.37–0.56)	0.48 (0.38–0.64)	0.549
Main pulmonary artery (median, IQR$_{25}$-IQR$_{75}$)	10.00 (9.05–11.65)	12.55 (11.40–14.15)	0.01
Newborn (n, %)	39 (63.9%)	8 (47.1%)	0.165
BAV (n, %)	45 (73.8%)	10 (58.8%)	0.146
Borderline left ventricle (n, %)	11 (18.0%)	1 (5.9%)	0.025
Recoarctation of the aorta (n, %)	11.0 (18.0%)	3.0 (17.6%)	0.72

DISCUSSION

As demonstrated in the majority of studies conducted in both pediatric and adult patients, BAV and CoA are the most prevalent in male individuals.[17–19] This finding is supported by the results of our study. The implications of the coexistence of these two conditions are not yet fully understood. The present study did not find statistically significant differences regarding the studied echocardiographic parameters between group A and group B. A review of the literature reveals that patients with BAV and CoA develop complications related to BAV at an earlier age than those with BAV alone, and the most common surgical intervention is for aortic regurgitation.[18]

The impact of surgical or interventional treatment of CoA on the development of valvular complications associated with BAV has not yet been widely investigated in the scientific literature. A limited number of studies have investigated the relationship between BAV-CoA and the occurrence of aortic stenosis or aortic regurgitation in pediatric patients.[19] The findings of these studies indicate that children with BAV-CoA exhibit a lower prevalence of aortic stenosis or aortic regurgitation compared to children with pure BAV, and a reduced incidence of valvular intervention. These findings suggest that the surgical or in-

terventional correction of CoA does not appear to have a detrimental effect on BAV among pediatric patients. Some studies in adults have shown that patients with BAV and associated CoA did not develop aortic valve complications over time.[18] In contrast, a study of patients with CoA found that BAV is a predictor of the development of recoarctation in patients with CoA.[20]

Although BAV is the most prevalent CHD, other common conditions in patients with CoA include atrial septal defect, ventricular septal defect, valvular aortic stenosis, aortic arch hypoplasia, and borderline left ventricle.[9,21]

The pathophysiological effects of this pathology are caused by the difference in blood pressure between the upper and lower portions of the body, which most commonly oscillates between 30 and 40 mmHg at rest.[22,23] Over the course of our study, blood pressure values exhibited a range of 20–25 mmHg. This complex process is affected by a number of factors, including the activity of baroreceptors, the presence of circulating catecholamines, the functioning of the angiotensin system, and residual lesions in the surgically corrected coarctation area.[9,24]

A variety of multiparametric diagnostic models have been described in the literature over the years, integrating different ultrasound signs for the detection of CoA in the neonatal period and infants. The I/D index and CSAI have

TABLE 9. Comparative subgroup analysis according to the I/D index

Variable	Low (≤ 0.64)	High (> 0.64)	p value
Age (months, median, IQR_{25}-IQR_{75})	0.00 (0.00–5.00)	0.00 (0.00–1.60)	0.314
Weight (kg, median, IQR_{25}-IQR_{75})	3.92 (3.40–5.43)	3.70 (3.00–5.50)	0.395
Length (cm, median, IQR_{25}-IQR_{75})	57.00 (53.00–64.50)	53.00 (51.50–62.50)	0.301
BSA (m², median, IQR_{25}-IQR_{75})	0.24 (0.21–0.30)	0.21 (0.21–0.29)	0.3
Ascending aorta (mm, median, IQR_{25}-IQR_{75})	8.36 (7.40–10.30)	7.00 (6.75–7.85)	0.026
Ascending aorta Z-score (median, IQR_{25}-IQR_{75})	−1.42 (−2.45 to 0.42)	−2.45 (−3.28 to −2.00)	0.015
Right innominate artery (mm, median, IQR_{25}-IQR_{75})	4.70 (4.09–5.80)	5.21 (4.21–5.80)	0.772
Distance between left common carotid artery and the left subclavian artery (mm, median, IQR_{25}-IQR_{75})	6.20 (4.40–7.34)	3.55 (3.17–3.80)	0.002
CSAI (median, IQR_{25}-IQR_{75})	0.95 (0.71–1.27)	1.21 (1.06–1.64)	0.148
Aortic isthmus (mm, median, IQR_{25}-IQR_{75})	2.60 (2.20–3.20)	3.60 (3.15–3.94)	0.004
Aortic isthmus Z-score (mean ± s.d.)	−5.30 ± 1.88	−3.53 ± 1.22	0.001
Descending aorta at diaphragm (mm, mean ± s.d.)	6.10 (5.00–7.20)	4.20 (4.00–4.85)	<0.001
I/D index (mean ± s.d.)	0.43 ± 0.12	0.80 ± 0.16	<0.001
BAV (n, %)	43 (70.5%)	7 (63.6%)	0.562
Aortic arch hypoplasia (n, %)	28 (45.9%)	6 (54.5%)	0.559
Borderline LV (n, %)	10 (16.4%)	2 (18.2%)	0.118
Recoarctation (n, %)	12.0 (19.7%)	1.0 (9.1%)	1
Newborn (n, %)	36 (59.0%)	7 (63.6%)	0.88
BAV (n, %)	45 (73.8%)	10 (58.8%)	0.146
Borderline left ventricle (n, %)	11 (18.0%)	1 (5.9%)	0.025
Recoarctation of the aorta (n, %)	11.0 (18.0%)	3.0 (17.6%)	0.72

demonstrated, over time, their sensitivity and specificity for CoA.[25] To the best of our knowledge, no studies have been conducted in our country on these parameters in infants older than 3 months of age or in cases of aortic recoarctation. Consequently, the mean value of the CSAI was 0.97, whereas that of the I/D index was 0.47 for the entire sample. Statistically significant results were obtained upon comparing CSAI values between newborns and the remaining children in the group (p < 0.035), whereas no statistically significant data was found for the I/D index. With regard to the presence or absence of BAV, no data with statistical significance were identified for the two indices. In light of these outcomes, it can be postulated that patients exhibiting lower CSAI values (≤1.5) have diminished absolute values in the ascending aorta and aortic isthmus, accompanied by augmented distances between the common carotid artery and the subclavian artery. The evaluation of the I/D index yielded comparable results.

Over time, several types of surgical corrections have been described: resection of the coarctation zone and termino-terminal reanastomosis, interposition of a vascular heterologous prosthesis, vascular heterologous patch plasty, widening aortoplasty with subclavian artery flap, or aorto-aortic bypass with vascular heterologous prosthesis. These procedures are generally performed by left lateral thoracotomy. Rao found operative mortality to be between 0% and 50% with a mean of 19%, whereas recoarctation rates ranged between 0% and 100% with a mean of 17%.[26] In our study, recoarctation of the aorta occurred in 20% of patients. Young age at the time of surgery, the presence of aortic arch hypoplasia, and short body length were identified as risk factors for recoarctation. Balloon angioplasty with or without stenting is the preferred intervention for adolescents and adults, being an effective treatment in 96–98% of cases. Rao compared the efficacy and safety of balloon angioplasty and surgical correction in infants < 3 months of age, concluding that balloon angioplasty may be an accepted alternative for the symptomatic treatment of CoA in this patient population.[26] With regard to secondary hypertension, 62.4% of patients exhibited this complication at the time of discharge. The precise mechanism underlying the development of hypertension following surgical correction remains unclear. Potential contributing factors may include aortic dysfunction, impaired baroreceptor function and left ventricular dysfunction.

CONCLUSION

The implications of the coexistence of CoA and BAV are not yet fully understood. Despite appropriate treatment,

patients with CoA have a reduced life expectancy and an increased risk of developing long-term cardiovascular complications, including those related to a BAV, recoarctation of the aorta, and secondary hypertension.

CONFLICT OF INTEREST

The authors declare no conflicts of interest.

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INFORMED CONSENT

Written informed consent was obtained from all subjects involved in the study.

DATA AVAILABILITY

Further data are available from the corresponding author upon reasonable request.

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