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# Assessment of anatomy of the aorta in patients with a coarctation of aorta

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## Abstract

**Background** Coarctation of the aorta (CoA) is a congenital heart disease characterized by the narrowing of the aorta, resulting in reduced blood flow to the body and increased pressure in the left ventricle. The pathophysiology of CoA is complex and involves several changes in the structure and function of the aorta. Recent studies have suggested that patients with CoA may have changes in the aortic wall beyond the site of the narrowing. Understanding these changes in the aorta is essential for optimizing the management of patients with CoA.

**Methods** Eighty-five patients who were diagnosed with an isolated coarctation of aorta (CoA) and underwent elective surgical repair—during the last 10 years were included in the study.

**Results** Eighty-five patients (62 males, 72.9%) with a median age of 7 years old (range from 1 month to 48 years old) underwent surgical repair of CoA during the last 10 years using 4 different methods of operation.

The study showed that more than a half (51.7%) of our patients with coarctation of the aorta had an enlargement of an ascending aorta, and *z*-score of ascending aortic size positively correlated with age, height, and weight, which means in patients with non-corrected CoA, ascending aorta tends to enlarge by time. In the present study, 31.8% of patients had an aortic arch hypoplasia, and aortic arch sizes *z*-score negatively correlated with ICU and hospital stay, which indicates that, patients with smaller aortic arch have poorer outcomes. Therefore, evaluating AAH before planning surgical repair is important for achieving better results. CT examination showed advantages in assessing aortic anatomy.

**Conclusions** It is suggested that an MSCT examination should be performed to take into consideration of current aortic anatomy, before planning the surgical correction of the aortic coarctation to achieve better results.

**Keywords** Congenital heart defects, Coarctation of aorta, Aortic anatomy, Enlargement of ascending aorta, Aortic arch hypoplasia, Computed tomography

## Background

Coarctation of the aorta (CoA) is a congenital heart disease characterized by the narrowing of the aorta, resulting in reduced blood flow to the body and increased pressure in the left ventricle. The most common location of CoA is just distal to the ligamentum arteriosum, which is a fibrous band that connects the left pulmonary artery and the descending aorta. CoA can present as an isolated lesion or in association with other cardiac defects, such as bicuspid aortic valve, ventricular septal defect, or hypoplastic left heart syndrome [1].

The pathophysiology of CoA is complex and involves several changes in the structure and function of the aorta. The most prominent change is the narrowing of the aorta, which can be diffuse or localized and can vary in severity from mild to severe. The narrowing of the

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aorta can lead to turbulent blood flow, which can cause endothelial damage, smooth muscle cell proliferation, and fibrosis. These changes can further exacerbate the narrowing and lead to complications such as hypertension, aortic stenosis, or aortic aneurysm [2].

Several imaging modalities are available to diagnose and monitor CoA, including echocardiography, cardiac magnetic resonance imaging (MRI), computed tomography (CT) scan, and cardiac catheterization. These modalities can provide detailed information about the location and severity of the aortic narrowing, as well as the presence of other cardiac defects.

Recent studies have suggested that patients with CoA may have changes in the aortic wall beyond the site of the narrowing. These changes, which include increased wall thickness, decreased distensibility, and altered collagen and elastin content, may be related to the hemodynamic stress caused by aortic stenosis and hypertension. Moreover, these changes may persist even after successful treatment of CoA, indicating that CoA has a long-lasting effect on the structure and function of the aorta [3].

Understanding these changes in the aorta is essential for optimizing the management of patients with CoA. For instance, knowledge of the altered aortic wall properties can help predict the risk of complications such as dissection or aneurysm formation, and guide decisions regarding timing and type of repair. Additionally, identifying the mechanisms underlying these changes can provide new targets for therapeutic interventions aimed at reducing the risk of long-term complications [4].

This paper aims to learn about the changes in the aorta in patients with CoA, focusing on the structural and functional alterations and their clinical implications. The paper will also discuss the underlying mechanisms of these changes and potential future directions for research and clinical management.

## Methods

A retrospective cohort study was performed. Medical records and CT scans were analyzed retrospectively and all relevant information, such as intraoperative procedures, intraoperative and postoperative complications, and CT scan measurements were acquired and collected in an Excel sheet to perform statistical analysis. All patients' information and CT scan measurements were collected in different Excel sheets to reduce performance and detection bias.

Clinical audits do not involve anything being done to patients beyond their normal clinical management and, therefore do not require patient consent or formal ethical review or approval. The present study was registered as a clinical audit, and all data were anonymized.

Eighty-five patients who were diagnosed with an isolated coarctation of aorta (CoA) and underwent elective surgical repair—during the last 10 years were included in the study.

Patient information in this research included results of preoperative medical examination with EchoCG and MSCT findings, postoperative patient status in the hospital with EcoCG results, and findings of medical examination at follow-up at 1 month and 1 year after operation with EcoCG.

Preoperative CT scans of patients were analyzed and measurements were performed using "Syngo.via ProtoNeo" software (Siemens Healthcare GmbH/Siemens Medical Solutions USA, 2018). All measurements were done as stated in the Society for Vascular Surgery guidelines and reporting standards [5]. The diameter of the aorta is measured in 5 parts, such as ascending aorta, aortic arch, isthmus (usually same with coarctation cite), descending aorta, and Ao in diaphragm level (Fig. 1). *Z*-scores for all measurements were found using the website: http://parameterz.blogspot.com/2008/09/z-scores-of-cardiac-structures.html. Body surface area (BSA) was calculated using the DuBois formula:

$$\text{BSA (m2)} = \sqrt{\frac{\text{height (cm)x weight}\left(\text{kg}\right)}{3600}}.$$

All statistical analyses were carried out with SPSS Statistics 22.0 software (IBM corporation and other(s) 1989, USA, 2013), and results were checked for homogeneity (Levene's test) and normal distribution (QQ-plot). Mean and standard deviation (SD) were used to summarize numerical variables with symmetric distribution, median, and inter-quantile range (IQR) for non-symmetric numerical variables. Correlation tests were calculated with the Pearson correlation coefficient. $P$ values $< 0.05$ were considered significant.

## Results

Eighty-five patients (62 males, 72.9%) with a median age of 7 years old (range from 1 month to 48 years old) underwent surgical repair of CoA during the last 10 years using 4 different methods of an operation: resection and end-to-end anastomosis, patch aortoplasty, interpositioning graft and resection with extended end-to-end anastomosis. One patient (1.2%) died 23 days after the operation, because of sepsis and lung issues.

All patients were diagnosed using echocardiography (EchoCG) and multi-slice computed tomography (MSCT) with contrast. EchoCG showed a pressure gradient on a coarctation site of the aorta (mean 53 mmHg ± 22.8), LV hypertrophy (in 54 patients, 63.5%), LV dilation (in 31 patients, 36.5%), and LV hypokinesis (ejection

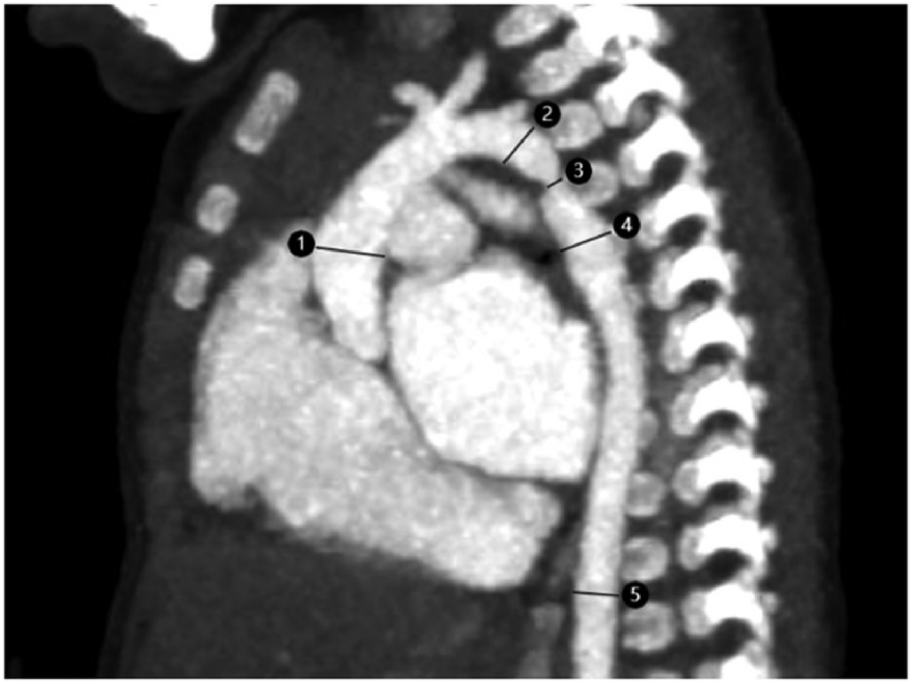


**Fig. 1** MSCT measurements of aortic morphology. Diameters: 1—ascending aorta, 2—aortic arch, 3—isthmus, 4—descending aorta, 5—Ao in diaphragm level

fraction of left ventricle was lower than 50%) (in 11 patients, 12.9%). Aortic regurgitation (AR) was assessed by EcoCG and the severity of AR was divided into 4 degrees from mild (1) to severe (4) (0 for without aortic regurgitation). It was revealed that in 74 patients (87%) aortic valve was without regurgitation (0°), 9 patients (10.6%) had 1st-degree AR, and 2 patients (2.3%) had 2nd-degree AR. There were no patients with 3rd and 4th-degree aortic regurgitation.

MSCT findings showed accurate sizes of the Aorta and its parts. All CT-scan measurements can be seen in Table 1.

Furthermore, enlargements (*Z*-score is greater than + 2.0) of ascending Ao (in 44 (51.7%) patients), aortic arch (in 6 patients (7%)), isthmus (in 1 patient, 1.2%), descending aorta (in 23 patients (27%)) and Ao in diaphragm level (in 19 patients (22.4%)) were revealed, while hypoplasia (*Z*-score is lower than − 2.0) of Ao Arch was evaluated in 27 patients (31.8%). The small size of the isthmus was analyzed in 84 patients (98.8%), who were diagnosed with the typical form of a CoA, and 2 patients were diagnosed with atypical CoA: 1 patient had small isthmus and descending Ao, and 1 patient's coarctation site was in a diaphragm level.

The correlation test showed that the EchoCG pressure gradient does not correlate to a *z*-score of the size of the aortic isthmus ($r = 0.276$, $p = 0.126$). Other results of the correlation test of *z*-scores can be seen in Table 2.

**Table 1** MSCT findings and z-scores of aortic anatomies

| Variables | Size (in mm) | | | *Z*-score | | |
|---|---|---|---|---|---|---|
| | Mean | Standard deviation (SD) | Range | Mean | Standard deviation (SD) | Range |
| Ascending aorta | 23.0 | ± 8.03 | 8.5–37.0 | + 2.19 | ± 1.64 | − 1.06–+ 5.49 |
| Aortic arch | 14.7 | ± 6.49 | 6–37.0 | − 0.743 | ± 1.85 | − 4.10–+ 3.54 |
| Isthmus | 5.51 | ± 3.66 | 1.2–21.0 | − 5.34 | ± 3.13 | − 13.2–+ 3.52 |
| Descending aorta | 15.5 | ± 5.99 | 5.5–31.0 | + 1.33 | ± 1.68 | − 2.54–+ 5.53 |
| Aorta in diaphragm level | 13.6 | ± 5.02 | 4.3–30.0 | + 0.925 | ± 2.05 | − 5.88–+ 6.12 |

**Table 2** Correlation of aortic anatomy with patient-related factors

| Variables | | Ascending aorta size *z*-score | | Aortic arch size *z*-score | | Aortic isthmus size *z*-score | | Descending aorta size *z*-score | | Ao in a diaphragm level size *z*-score | |
|---|---|---|---|---|---|---|---|---|---|---|---|
| | | *r* | *p* | *r* | *p* | *r* | *p* | *r* | *p* | | *p* |
| Age | | .373 | .018* | .335 | .035* | .058 | .721 | .198 | .221 | .021 | .897 |
| Height | | .381 | .026* | .266 | .128 | .035 | .844 | .047 | .798 | − .065 | .715 |
| Weight | | .354 | .025* | .264 | .100 | .053 | .744 | .178 | .272 | − .006 | .971 |
| BMI | | .367 | .544 | .232 | .707 | .935 | .020* | .897 | .039* | .542 | .345 |
| EchoCG findings | Pressure gradient on a coarctation cite | .249 | .169 | .214 | .240 | .276 | .126 | .039 | .831 | − .257 | .156 |
| | Ao regurgitation | .152 | .194 | − .066 | .576 | − .187 | .108 | − .040 | .731 | .185 | .113 |
| | LV hypertrophy | .180 | .268 | .118 | .469 | − .128 | .432 | − .016 | .922 | .002 | .991 |
| | LV dilation | .002 | .988 | .077 | .638 | .245 | .127 | .172 | .290 | − .042 | .797 |
| | LV hypokinesis | .078 | .630 | .130 | .423 | − .076 | .643 | − .043 | .791 | .053 | .742 |
| | LV Ejection fraction | .084 | .605 | − .071 | .662 | − .160 | .324 | .063 | .701 | .138 | .355 |
| Operation time | | .266 | .097 | .307 | .054 | .074 | .652 | − .171 | .292 | − .129 | .428 |
| Ao cross-clamp time | | .050 | .784 | .202 | .266 | .405 | .021* | − .284 | .115 | − .173 | .344 |
| ICU stay | | -.035 | .841 | − .355 | .034* | .135 | .416 | − .144 | .376 | − .107 | .511 |
| Hospital stay | | .099 | .545 | − .343 | .03* | .055 | .737 | − .082 | .615 | − .110 | .501 |

*r* Pearsons value, correlation coefficient; *p p* value

*$p < .05$, **$p < .01$, ***$p < .001$

Besides that, some *Z*-scores are correlated with each other (Table 3).

## Discussion

Coarctation of the aorta (CoA) is a congenital heart defect characterized by a narrowing of the thoracic aorta that obstructs blood flow to the lower part of the body. Multislice computed tomography (MSCT) is a non-invasive imaging modality that provides accurate and detailed images of the cardiovascular system, including the aorta and its branches. MSCT has been increasingly used in the diagnosis and follow-up of patients with CoA [6]. MSCT offers several advantages over other imaging techniques such as echocardiography, magnetic resonance imaging (MRI), and catheter angiography, including its ability to provide high spatial resolution and to visualize the aorta in three dimensions (3D) [7].

To evaluate the results, we used *z*-score for sizes of different parts of an aorta and correlated these findings with other patient-related factors and results of EchoCG.

Current literature shows, a range of MSCT findings in patients with CoA, including the exact location and severity of the coarctation, the presence of collateral vessels, and abnormalities of the aortic arch [8, 9].

**Table 3** Correlation of aortic anatomy with each other

| | Ascending aorta size *Z*-score | | Aortic arch size *z*-score | | Aortic isthmus size *z*-score | | Descending aorta size *z*-score | | Ao in a diaphragm level size *z*-score | |
|---|---|---|---|---|---|---|---|---|---|---|
| | *r* | *p* | *r* | *p* | *r* | *p* | *r* | *p* | *r* | *p* |
| Ascending aorta size *z*-score | | | | | | | | | | |
| Aortic arch size *z*-score | .660 | < .001*** | | | | | | | | |
| Aortic isthmus size *z*-score | .137 | .398 | .422 | .007 ** | | | | | | |
| Descending aorta size *z*-score | .111 | .494 | .254 | .114 | .025 | .880 | | | | |
| Ao in a diaphragm level size *z*-score | − .143 | .378 | − .141 | .386 | − .345 | .029* | .469 | .002* | | |

*r* Pearsons value, correlation coefficient; *p p* value

*$p < .05$, **$p < .01$, ***$p < .001$

There has been a recent increase in the number of studies comparing MSCT with TTE in the diagnosis of congenital CoA and accompanying malformations. Xu J et al. observed a series of 40 pediatric patients aged < 4 years with suspected CoA who underwent prospective ECG-triggered high-pitch CT angiography and TTE. Their results showed that the sensitivity, specificity, positive predictive value, negative predictive value, and overall diagnostic accuracy of CT in the evaluation of complex CoA were 92.37%, 98.51%, 97.32%, 93.57%, and 96.25%, respectively. For a total of 80 extra-cardiac anomalies, the sensitivity (98.8%, 79/80) of CT was greater than that of TTE (62.5%; 50/80). On the contrary, for 38 cardiac anomalies, the sensitivity (78.9%, 30/38) of CT was lower than that of TTE (100%; 38/38) [10].

In our study, we did not aim to compare these two methods; however, our results showed that the pressure gradient on the coarctation site of the Aorta did not correlate with the *z*-score of the coarctation site, which means TTE does not show enough evidence according to detecting the severity of coarctation. On the other hand, TTE is a non-invasive and fast method and does not require sedation of the patients. Therefore, we suggest that TTE should be used for primary diagnostics, especially to detect LV function and the presence of CoA, but we consider that identifying aortic anatomy before the surgical procedure using MSCT is crucial.

Despite several reports about enlargement of ascending aorta in patients with coarctation of aorta, the cause of this enlargement is still unclear and usually considers the influence of arterial hypertension. Our study showed that more than a half (51.7%) of our patients with coarctation of the aorta had an enlargement of an ascending aorta, and *z*-score of ascending aortic size positively correlated with age, height, and weight, which means in patients with non-corrected coarctation ascending aorta tends to enlarge by time (Fig. 2).

Mechanical wall stress is considered one of the possible reasons for aortic aneurysm formation and rupture [11], and several factors can affect mechanical wall stress, such as aortic wall stiffness, locoregional pressure, anatomical characteristics of the aorta, and others

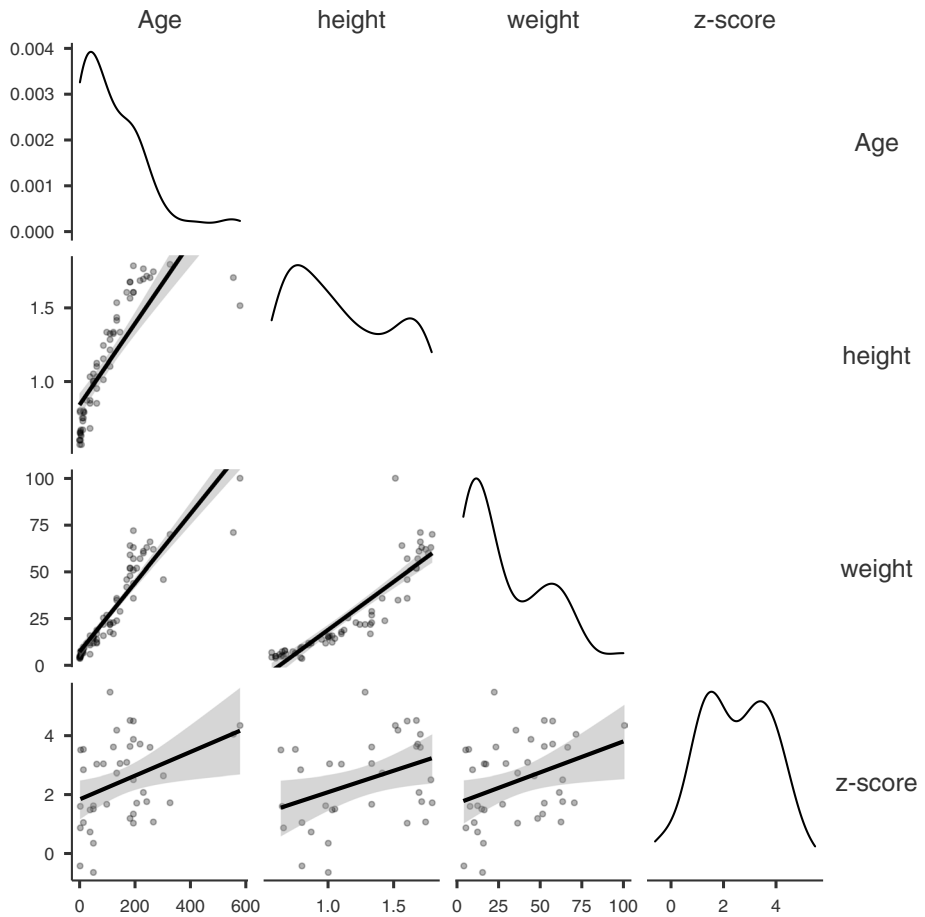


**Fig. 2** Correlation of *z*-score of ascending aortic size with age, height, and weight

[12]. The findings of this study suggest that coarctation of the aorta may affect mechanical wall stress and can be a potential risk for aortic enlargement and later aortic aneurysm formation. Currently, it is not feasible to assess aortic wall stress; however, coarctation of the aorta should be considered as a potential risk factor for ascending aortic aneurysm formation. To decrease this risk, it is suggested that, coarctation of the aorta should be repaired during early childhood.

Some scientific reports showed that the bicuspid aortic valve can cause to development of aortic regurgitation and enlargement of the ascending aorta, especially in patients with CoA [13, 14]. In our research, a non-regurgitated aortic valve was revealed in 87%, and severe AR was not detected. AR did not show a correlation with the *z*-score of ascending aorta or *z*-scores of other parts of the aorta. However, AR was positively correlated with age ($r = 0.408$, $p = 0.001$), height ($r = 0.282$, $p = 0.004$), and weight ($r = 0.299$, $p = 0.002$). *T* test showed that the diameter of ascending aorta in the AR group (mean 21.92 mm ± 9.25) and non-AR group (mean 16.31 mm ± 8.26) differed significantly ($p = 0.032$), while *z*-scores of ascending aortas in these two groups were not significantly different (2.10 ± 1.33 and 1.67 ± 1.32, respectively, $p = 0.294$). This suggests that an association between ascending aortic enlargement and regurgitation of the aortic valve cannot be confirmed, and AR tends to develop over time.

Another clinically important measurement using MSCT is aortic arch size. CoA combined with aortic arch hypoplasia (AAH) is a complex form of CHD. Without timely and effective treatment, patients with this combined disorder always have a poor prognosis. Conte et al. reported that the early appearance of clinical symptoms correlated strongly with severe conditions among newborns with CoA. In addition, up to 81% of these newborns had CoA combined with AAH [15]. For now, there are still no definite diagnostic criteria for AAH. In this study, the diagnostic criteria of AAH were based on the CHD database. According to these criteria, AAH is present if the AA diameter between the innominate artery and the left common carotid artery is < 60% of the ascending aorta diameter if the AA diameter between the left common carotid artery and the left subclavian artery is < 50% of the ascending aorta diameter, or if the diameter of the aortic isthmus is < 40% of the ascending aorta diameter [16]. However, the limitation of this measurement is the enlarged ascending aorta, which is common in patients with CoA. Therefore, we suggest that to evaluate AAH, the z-score for aortic arch size should be calculated, and lower than − 2.0 should be considered as an AAH.

In the present study, 31.8% of patients had an aortic arch hypoplasia, and aortic arch sizes *z*-score negatively correlated with ICU and hospital stay, which indicates that, patients with smaller aortic arch have poorer outcomes. Therefore, evaluating AAH before planning surgical repair is important for achieving better results.

Assessing the severity, location, and length of the coarctation site is another important measurement. In our study, 3 patients had an atypical form of coarctation of the aorta, which was detected using MSCT examination. For these patients, we used an interposition graft to repair the defect. Besides that, the *z*-score for isthmus size showed a positive correlation with an aortic cross-clamp time. Thus, patients who had wider isthmus had an atypical form of CoA, and repairing these types of defects required more time.

Enlargements of the descending aorta (in 27% of patients) and the aorta at a diaphragm level (22.4%) can be explained with post-stenotic dilatation of the aorta. The etiology of post-stenotic aortic dilatation remains controversial. It may be due to hemodynamic factors caused by a stenotic site, involving high velocity and turbulent flow downstream of the stenosis, or due to intrinsic pathology of the aortic wall. This may involve an abnormality in the process of extracellular matrix remodeling in the aortic wall including inadequate synthesis, degradation, and transport of extracellular matrix proteins [17]. The negative correlation of these *z*-scores with isthmus size, and strong positive correlation to each other, can be proof of that hypothesis.

The study has several limitations, the most important limitation is retrospective design, because of retrospective some information, such as diameters of ascending aorta, aortic arch, and descending aorta, and bicuspidity of aortic valve could not be found in pre-operative EchoCG results, which could help to assess the anatomy of aorta and role of CT scan. Therefore, we suggest that future work in this area should look at differences in parameters that will be obtained by two methods: EcoCG and MSCT to assess these diagnostic tools on revealing aortic anatomy, especially according to age groups. Another future direction can be learning the influence of the bicuspid aortic valve on aortic anatomy and its changes over time in the bicuspid and tricuspid aortic valve groups.

## Conclusions

The present study proved that coarctation of the aorta has a huge impact on the anatomy of the ascending aorta, aortic arch, and descending aorta. CT examination showed advantages in assessing aortic anatomy. It is suggested that an MSCT examination should be performed to take into consideration of current aortic anatomy, before planning the surgical correction of the aortic coarctation to achieve better results.

**Abbreviations**

| | |
|---|---|
| CoA | Coarctation of aorta |
| ICU | Intensive care unit |
| AAH | Aortic arch hypoplasia |
| CT | Computed tomography |
| MSCT | Multislice computed tomography |
| MRI | Magnetic resonance imaging |
| BSA | Body surface area |
| SD | Standard deviation |
| IQR | Inter-quantile range |
| EchoCG | Echocardiography |
| LV | Left ventricle |
| Ao | Aorta |
| AR | Aortic regurgitation |
| TTE | Trans-thoracic echocardiography |
| CHD | Congenital cardiac defects |
| AA | Aortic arch |

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**Authors' contributions**
BT analyzed and interpreted the patient data, wrote the paper, HA performed statistical analysis, and BH and NI collected the patient data. All authors read and approved the final manuscript.

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**Availability of data and materials**
All data were collected in an Excel sheet and can be provided if necessary.

## Declarations

**Ethics approval and consent to participate**
The ethics committee of cardio-vascular surgery of the Center of Surgery in Tashkent approved this study (REC number: 46522). The research does not require consent because of retrospective character.

**Consent for publication**
Not applicable.

**Competing interests**
The authors declare that they have no competing interests.

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## Publisher's Note