Astana Medical Journal
e2790-1203

https://doi.org/10.54500/2790-1203-2025-6-125-amj002

Agreement between Z-score and an arch-to-ascending aorta ratio derived from CT angiography for detection of aortic arch hypoplasia

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Received: 15.08.2025
Accepted: 22.10.2025
Published: 29.12.2025

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Citation: Astana Medical Journal, 2025, 125 (6), amj002.

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Abstract

Background. Aortic arch hypoplasia (AAH) is an important congenital cardiovascular abnormality that requires accurate anatomical characterization for appropriate surgical planning. Computed tomography angiography (CTA) provides high-resolution measurements of the ascending aorta and transverse aortic arch; however, different diagnostic criteria may classify hypoplasia inconsistently.

Objective. This study aimed to compare the agreement between a Z-score-based definition of AAH and a ratio-based morphologic criterion derived from CTA measurements.

Methods. This retrospective study included 48 pediatric patients with clinically confirmed AAH who underwent CTA. The ascending aorta diameter, transverse aortic arch diameter, and the arch-to-ascending aorta (Arch/AAo) ratio were obtained from multiplanar reformatted CTA images. Hypoplasia was defined using two approaches: (1) Z-scores (≤ −2), and (2) a ratio-based criterion (Arch/AAo < 0.50). Summary statistics were computed for all measurements, and agreement between methods was assessed using percent agreement, Cohen's κ, and McNemar's test.

Results. The mean ascending aorta diameter was 0.95 ± 0.46 cm, the mean transverse arch diameter was 0.50 ± 0.27 cm, and the median Arch/AAo ratio was 0.50 (IQR 0.43-0.65). The Z-score method classified 47 of 48 patients (97.9%) as hypoplastic, whereas the ratio criterion identified 18 patients (37.5%) as hypoplastic. Agreement between methods was 44.2%, with a Cohen's κ of 0.03, indicating minimal concordance beyond chance. McNemar's test demonstrated significant disagreement between classifications ($\chi^2 = 22.04$).

Conclusion. The Z-score and Arch/AAo ratio methods differ substantially in how they classify AAH. While Z-scores incorporate normative size adjustment, the ratio criterion reflects anatomical proportionality and identifies a more selective subset of patients with marked transverse arch narrowing. Clinicians should recognize these methodological differences when assessing AAH and selecting criteria for diagnosis or surgical decision-making.

Keywords: aortic arch hypoplasia, congenital heart defects, computed tomography angiography, Z-score, aortic ratio, pediatric cardiovascular imaging, aortic measurements, diagnostic agreement.

1. Introduction

Congenital anomalies of the aortic arch are relatively uncommon and may coexist with other congenital cardiovascular disorders [1, 2]. In most cases, these anomalies are detected incidentally on imaging in otherwise asymptomatic patients [3]. An important exception occurs when the aberrant arch configuration forms a complete vascular ring, encircling the trachea and esophagus and potentially producing compressive symptoms [4].

Aortic arch hypoplasia (AAH) is defined by comparing the external diameter of each arch segment with that of the ascending aorta, which is assumed to represent normal caliber [5]. Based on established criteria, the proximal transverse arch is considered hypoplastic when its external diameter measures <60% of the ascending aorta (AAo), the distal transverse arch when <50% [6, 7], and the aortic isthmus when <40% of the AAo diameter [8]. AAH may occur as an isolated abnormality or in association with other aortic lesions that impede systemic outflow, such as coarctation and interruption of the aorta [9]. It may also coexist with intracardiac defects, including atrial septal defect, ventricular septal defect, or patent ductus arteriosus [10].

Computed tomography angiography (CTA) is a non-invasive imaging modality that enables accurate detection and characterization of aortic arch anomalies through high-resolution visualization of anatomical relationships, advanced post-processing techniques (Volume Rendering, Maximum Intensity Projection, and Multiplanar Reformation), and the ability to identify associated congenital abnormalities [11]. The high spatial resolution of CTA, combined with its capacity to evaluate extracardiac structures such as the great vessels, makes it the preferred modality for generating three-dimensional models in congenital heart disease [12, 13] *(Figure 1)*.

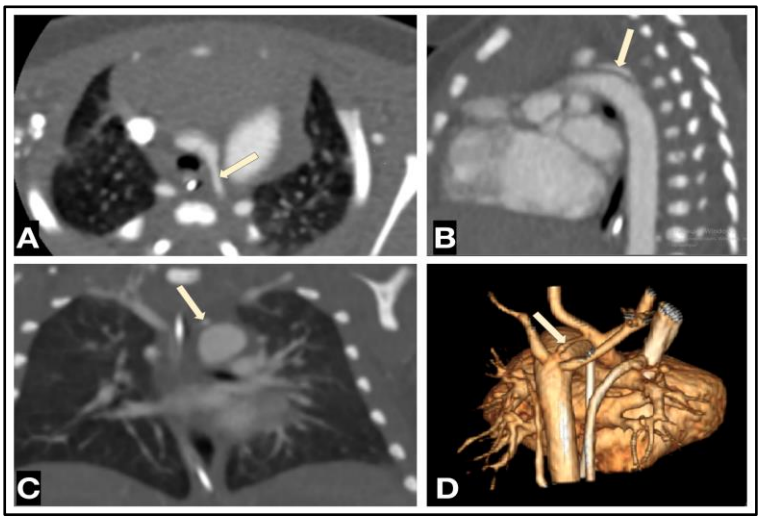

Figure 1 - CTA multiplanar images of a 5-day-old patient with AAH. (A) Axial plane, (B) sagittal plane, and (C) coronal plane demonstrate the narrowed transverse aortic arch, indicated by the yellow arrow. (D) Three-dimensional volume-rendered reconstruction provides an overview of the aortic arch anatomy, with the hypoplastic segment similarly highlighted

Despite the central role of CTA imaging in the evaluation of congenital aortic arch pathology, there is no consensus regarding the optimal criterion for defining AAH, and different measurement approaches may classify the same anatomy inconsistently. In particular, Z-score-based assessment [14, 15] and arch-to-ascending aorta (Arch/AAo) ratio-based morphological criteria are both used in clinical practice, yet their level of agreement has not been adequately examined. Therefore, this study aimed to compare these two commonly employed methods: Z-scores and the Arch/AAo diameter ratio using CTA-derived measurements in a cohort of patients with clinically confirmed AAH. By assessing the concordance between these approaches, we sought to clarify their diagnostic alignment and highlight potential implications for clinical evaluation and surgical planning.

2. Material and methods

Study design and population

This retrospective study included 48 consecutive pediatric patients with a confirmed clinical diagnosis of AAH who underwent CTA as part of their diagnostic evaluation at the Heart Center of the University Medical Center in Astana, Kazakhstan, between 2020 and 2023. Inclusion criteria were patients with available CTA imaging of sufficient quality to allow precise measurement of both the AAo and the aortic arch. Patients with incomplete CTA datasets, nondiagnostic image quality, or prior aortic surgery were excluded. All diagnoses were established by a multidisciplinary team consensus consisting of pediatric cardiologists, cardiothoracic surgeons, and radiologists.

Written informed consent was obtained from the legal representatives of pediatric patients for publication and any accompanying images. All procedures performed in studies involving human participants were in accordance with the ethical standards of the institutional and/or national research committee and with the 1964 Helsinki Declaration and its later amendments or comparable ethical standards. The study was approved by the Bioethics Committee of the Heart

Center, University Medical Center, Astana, Kazakhstan (21 Jan 2022/No. 01-110/2022).

Imaging acquisition

CTA examinations were performed using a Siemens Somatom Definition AS 64-slice scanner. Prospective ECG gating was applied to minimize cardiac motion artifacts. Intravenous iodinated contrast medium (Ultravist 370; 1.5-2.0 mL/kg) was administered using a dual-head injector at an infusion rate of 0.5-2.0 mL/s. Bolus tracking was performed with the region of interest positioned in the AAo and an acquisition trigger threshold of 100 Hounsfield units. Axial images were reconstructed with a slice thickness of 0.6 mm and a reconstruction increment of 0.1 mm. Multiplanar reformations were generated using syngo.via (Siemens, Germany) to obtain measurements perpendicular to the vascular axis.

Aortic diameter measurements

Z-score method [14]: the Z-score value reported in the CTA report (Heart Center institutional pipeline) was used to determine hypoplasia. Hypoplasia is defined as $Z \leq -2$. Z-scores were calculated by the CTA reporting software, normalized to body surface area.

AAH was defined using the Arch/AAo diameter ratio:

$$\frac{Aortic\ \ arch\ \ diameter}{Ascending\ \ aorta\ \ diameter} < 0.5.$$

Aortic arch diameter/Ascending aorta diameter<0.5.

This threshold is consistent with established radiologic and surgical criteria for clinically significant transverse arch hypoplasia [15].

Statistical Analysis

Statistical analyses were performed using Stata version 18.0 (STATA, StataCorp, Texas, US). Continuous variables were summarized using mean, standard deviation (SD), median, and interquartile range (IQR). Categorical variables were expressed as counts and percentages, with comparisons conducted using the Chi-square or Fisher's exact test, as appropriate. Normality was assessed using the Shapiro-Wilk test. Agreement between the Z-score method and the ratio-based method was evaluated using overall percent agreement, Cohen's κ statistic, and McNemar's test based on a 2×2 contingency table. A p-value < 0.05 was considered statistically significant.

3. Results

Patient cohort

A total of 48 patients with clinically confirmed AAH were included. After cleaning and standardization of CTA measurements, complete aortic diameter data (AAo and aortic arch) were available in all analyzable cases, and these were used for the Arch/AAo ratio calculation. The distribution of CTA-derived diameters is presented in *Table 1*.

Table 1 - Descriptive statistics of aortic measurements

Measurement	Mean	SD	Median	IQR	Min	Max
AAo	0.946	0.458	0.80	0.70-1.00	0.40	3.00
Arch	0.497	0.269	0.40	0.385-0.50	0.18	1.70
Ratio	0.539	0.154	0.50	0.43-0.65	0.30	1.00
Z-score (CTA)	-4.149	1.748	-3.965	-4.752- -2.950	-9.40	-1.98

Arch-to-ascending aorta diameter ratio

The Arch/AAo ratio was calculated in all patients with complete aortic measurements. The distribution of ratio values *(Figure 1)* demonstrated substantial anatomical variability, with a subset of patients exhibiting markedly reduced ratios consistent with pronounced transverse arch narrowing. Histogram plots of the AAo diameter, transverse aortic arch diameter, and the resulting Arch/AAo ratio illustrate the variability within the cohort and highlight the morphological differences captured by this proportional metric.

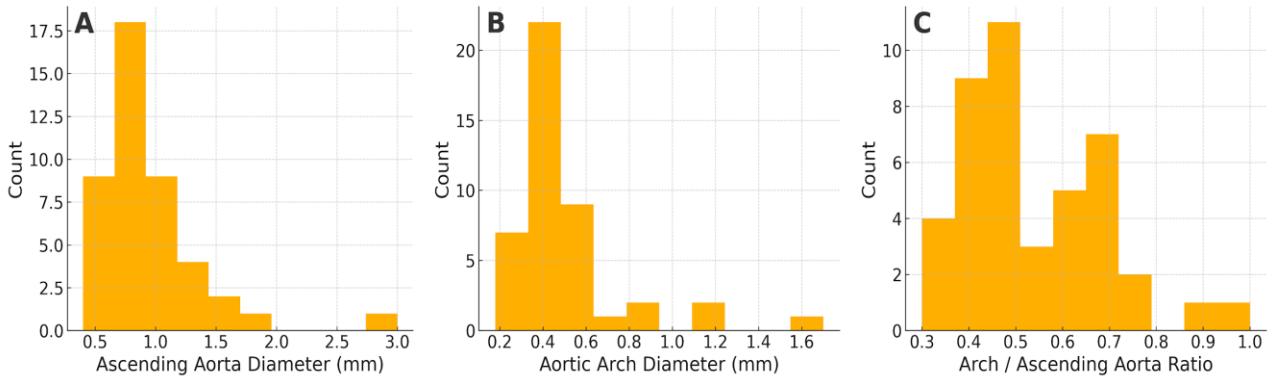

Figure 2 - Distribution of AAo, aortic arch, and Arch/AAo ratio measurements in the study cohort. The panel presents histograms of (A) AAo diameter, (B) transverse aortic arch diameter, and (C) the Arch/AAo ratio. All measurements were obtained from CTA in patients with complete aortic diameter data included in the final analysis

Agreement between the Z-score and the arch-to-ascending aorta ratio

An agreement analysis between the Z-score method and the ratio-based criterion was performed in all 48 patients *(Table 2)*. Using the <0.50 threshold, 18 patients were classified as hypoplastic by the ratio method. Based on available and clinically derived Z-score classifications, 47 patients were categorized as hypoplastic. The agreement analysis showed 18 concordant hypoplastic classifications and 25 discordant classifications, with one case classified as negative by both methods. Overall agreement was 44.2%. Cohen's κ was 0.03, and McNemar's test yielded a χ^2 value of 22.04.

Table 2 - Agreement between methods

Comparison	n concordant hypoplasia	n discordant	κ (95% CI)	McNemar p
Z-score vs Ratio	22	25	0.03	22.04

Associated cardiovascular anomalies

Chi-square and Fisher's exact tests were applied to assess the clinical characteristics of patients with AAH and their associated chromosomal and cardiovascular anomalies, which were diagnosed using echocardiography and CTA. The results of these analyses are presented in *Table 3*.

Table 3 - Clinical data of 48 patients with AAH and their associated cardiovascular anomalies

Characteristics	Options	Number (Percentage)
Imaging modalities	64-slice CTA	48 (100.0%)
	Echocardiography	48 (100.0%)
Aortic arch side	Left	46 (95.8%)
	Right	2 (4.2%)
Chromosome abnormalities	Down syndrome	2 (4.2%)
Combined cardiovascular anomalies	Ventricular septal defect	33 (68.8%)
	Atrial septal defect	21 (43.8%)
	Atrioventricular septal defect	5 (10.4%)
	Coarctation of the aorta	28 (58.3%)
	Patent ductus arteriosus	33 (68.8%)
	Patent foramen ovale	19 (39.6%)
	Aortopulmonary window	1 (2.1%)
	Transposition of the great arteries	8 (16.7%)
	Bicuspid aortic valve	7 (14.6%)
	Ebstein's anomaly	1 (2.1%)
	Taussig-Bing anomaly	3 (6.3%)
Myocardial hypertrophy	Left	8 (16.7%)
	Right	1 (2.1%)
	Both	1 (2.1%)

4. Discussion

The comparison between the ratio-based method and the Z-score classification demonstrated substantial variation in how each approach identifies AAH. The ratio method, which reflects the proportional relationship between the aortic arch and the AAo [17], classified a smaller proportion of patients as hypoplastic, whereas the Z-score method identified nearly all patients as hypoplastic. This resulted in a low level of agreement between the two approaches. Cohen's κ was near zero, indicating minimal concordance beyond chance, and McNemar's test showed significant directional disagreement *(Table 2)*, suggesting that the two methods categorize patients differently in a non-random manner.

These findings indicate that while CTA remains essential for evaluating aortic arch anatomy [18], notable methodological differences exist between the size-adjusted Z-score assessment and the morphology-based ratio criterion. The ratio method emphasizes structural disproportionality between aortic segments [19], while Z-scores rely on deviation from normative pediatric reference data [20]. As a result, the ratio approach appears to function as a more restrictive anatomical measure, identifying only those patients with pronounced narrowing of the transverse aortic arch.

The ratio has been widely adopted as a diagnostic criterion in prior research. Kiraly et al. identified an empiric threshold of 0.5 for differentiating normal from hypoplastic arches based on the distribution of Arch/AAo ratios. Most associated congenital anomalies were similarly distributed across both groups, except for atrial septal defect, which was consistently more frequent among patients with a hypoplastic arch [21]. In their case report describing endovascular treatment of recurrent aortic hypoplasia and coarctation in a 15-year-old patient, Rhodes et al. defined a hypoplastic aortic arch as an Arch/AAo diameter ratio of <0.5 [22]. Despite its widespread use, the definition and management of aortic hypoplasia, especially in relation to coarctation, remain areas of ongoing research and clinical debate [23-25].

This study has several limitations. Its retrospective design and the absence of standardized measurement acquisition may introduce selection and measurement bias. Z-scores were inconsistently documented and could not be recalculated from raw data, reducing uniformity in the reference standard. The relatively small cohort and the imbalance between hypoplastic and non-hypoplastic classifications also constrained the robustness of agreement statistics. Larger prospective studies with standardized measurements and recalculated Z-scores are needed to validate these findings.

5. Conclusion

This study evaluated two commonly used approaches for identifying AAH: Z-score assessment and a morphologic ratio-based criterion derived from CTA measurements. The ratio method identified a smaller subset of patients as having a hypoplastic arch, resulting in limited agreement with Z-score classification. These findings emphasize that the two methods reflect distinct aspects of aortic arch anatomy: Z-scores incorporate normative size adjustment, whereas the ratio criterion captures relative structural narrowing. When interpreting imaging findings or planning surgical management, clinicians should be aware of these methodological differences, as the selected definition may influence which patients are categorized as having clinically significant AAH. Further research incorporating standardized measurement protocols and larger cohorts may help refine the optimal approach for consistent and clinically meaningful identification of arch hypoplasia.

Funding

The authors declare that no funds, grants, or other support were received during the preparation of this manuscript.

Conflict of Interest

The authors declare that the research was conducted in the absence of any commercial or financial relationships that could be construed as a potential conflict of interest.

Author Contributions

Conceptualization: Moldakhanova Zh.A.; Methodology: Moldakhanova Zh.A.; Formal analysis and investigation: Nurmakhan Zh. ZH, Bastarbekova L.A.; Writing - original draft preparation: Moldakhanova Zh.A., Zholshybek N. ZH; Writing - review and editing: Dautov T.B., Zholshybek N. ZH; Visualization: Zh.S. Abdrakhmanova; Resources: Dautov T.B.; Supervision: Rakhimzhanova R.I.

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Қолқа доғасының гипоплазиясын анықтау үшін КТ-ангиография деректерінен алынған Z-көрсеткіш пен қолқа доғасының өрлемелі қолқаға қатынасының арасындағы келісімділік

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Түйіндеме

Кіріспе. Қолқа доғасының гипоплазиясы (ҚДГ) хирургиялық тактиканы дұрыс жоспарлау үшін дәл анатомиялық бағалауды талап ететін маңызды туа біткен жүрек-қантамыр аномалиясы болып табылады. Компьютерлік томография ангиографиясы (КТ-ангиография) өрлемелі қолқаның және көлденең қолқа доғасының өлшемдерін жоғары кеңістіктік айырымдылықпен анықтауға мүмкіндік береді; алайда әртүрлі диагностикалық критерийлер гипоплазияны әрқалай жіктеуі мүмкін.

Зерттеудің мақсаты. ҚДГ анықтауда қолданылатын КТ-ангиография деректерінен алынған Z-көрсеткішке негізделген тәсіл және арақатынасқа негізделген морфологиялық критерийдің өзара келісімділігін бағалау.

Әдістері. Бұл ретроспективті зерттеуге клиникалық түрде ҚДГ расталып КТ-ангиография жасалған 48 педиатриялық науқас енгізілді. КТ-ангиография суреттерінің көпкеңістікті реконструкциясынан өрлемелі қолқаның диаметрі, көлденең қолқа доғасының диаметрі және доға мен өрлемелі қолқаның арақатынасы (Arch/AAo) алынған. Гипоплазия екі тәсілмен анықталды: (1) Z-көрсеткіші ≤ –2 және (2) Arch/AAo қатынасы < 0,50. Барлық өлшемдер үшін сипаттамалық статистика жүргізілді, ал әдістер арасындағы келісімділік пайыздық сәйкестік, Коэн к коэффициенті және Мак-Немар тестін қолдану арқылы бағаланды.

Нәтижесі. Өрлемелі қолқаның орташа диаметрі 0,95 ± 0,46 см, көлденең доғаның орташа диаметрі 0,50 ± 0,27 см, ал Arch/AAo қатынасының медианасы 0,50 (IQR 0,43–0,65) болды. Z-көрсеткіш тәсілі 48 науқастың 47-сін (97,9%) гипоплазия деп жіктесе, қатынас критерийі 18 науқасқа (37,5%) гипоплазия диагнозын қойды.

Әдістердің жалпы сәйкестігі 44,2% болды, ал Коэн к коэффициенті 0,03 құрап, кездейсоқ сәйкестіктен жоғары минималды келісімділікті көрсетті. Мак-Немар тесті жіктеулер арасында айтарлықтай айырмашылықты анықтады ($\chi^2 = 22,04$).

Қорытынды. Z-көрсеткішке және Arch/AAo қатынасына негізделген әдістер ҚДГ жіктелуінде айтарлықтай айырмашылық көрсетеді. Z-көрсеткіштері өлшемдердің қалыпты өлшемнен ауытқуын ескерсе, қатынас критерийі анатомиялық пропорцияны сипаттайды және көлденең доғаның айқын тарылуы бар науқастардың тар ауқымын анықтайды. ҚДГ-ны бағалау және диагностикалық не хирургиялық шешім қабылдау барысында клиницистер бұл әдістемелік айырмашылықтарды ескеруі тиіс.

Түйін сөздер: қолқа доғасының гипоплазиясы, туа біткен жүрек ақаулары, КТ-ангиография, Z-көрсеткіш, аорталық қатынас, педиатриялық жүрек-қантамыр визуализациясы, қолқа өлшемдері, диагностикалық келісімділік.

Согласованность между Z-оценкой и отношением диаметра дуги аорты к восходящей аорте, рассчитанным по данным КТ-ангиографии, для выявления гипоплазии дуги аорты

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Резюме

Введение. Гипоплазия дуги аорты (ГДА) является важной врождённой сердечно-сосудистой аномалией, требующей точной анатомической оценки для планирования хирургического вмешательства. Компьютерная томографическая ангиография (КТ-ангиография) обеспечивает высокое пространственное разрешение для измерения восходящей аорты и поперечной дуги аорты; однако применение различных диагностических критериев может приводить к неоднозначной классификации гипоплазии.

Цель исследования. Оценить согласованность между определением ГДА на основе Z-оценки и морфологическим критерием, основанным на соотношении диаметров, рассчитанным по данным КТ-ангиографии.

Методы. В ретроспективное исследование были включены 48 педиатрических пациентов с клинически подтверждённой ГДА, которым была выполнена КТ-ангиография. Из многоплоскостных реконструкций были получены параметры: диаметр восходящей аорты, диаметр поперечной дуги аорты и отношение дуги к восходящей аорте (Arch/AAo). Гипоплазия определялась двумя способами: (1) Z-показатель ≤ –2 и (2) критерий

отношения Arch/AAo < 0,50. Для всех измерений были рассчитаны описательные статистики, а согласованность методов оценивалась с использованием процента совпадений, коэффициента к Коэна и критерия Мак-Немара.

Результаты. Средний диаметр восходящей аорты составил 0,95 ± 0,46 см, средний диаметр поперечной дуги — 0,50 ± 0,27 см, а медиана отношения Arch/AAo — 0,50 (IQR 0,43–0,65). Метод Z-оценки классифицировал 47 из 48 пациентов (97,9%) как имеющих гипоплазию, тогда как критерий отношения выявил гипоплазию у 18 пациентов (37,5%). Общая согласованность методов составила 44,2%, а коэффициент к Коэна — 0,03, что свидетельствует о минимальном совпадении сверх уровня случайности. Критерий Мак-Немара показал существенные расхождения между классификациями ($\chi^2 = 22,04$).

Заключение. Методы, основанные на Z-показателях и отношении Arch/AAo, существенно различаются в классификации ГДА. Z-показатели учитывают отклонение размеров от нормативов, тогда как критерий отношения отражает анатомическую пропорциональность и выделяет более узкую подгруппу пациентов с выраженным сужением поперечной дуги. При оценке ГДА и выборе диагностического или хирургического подхода клиницистам следует учитывать эти методологические различия.

Ключевые слова: гипоплазия дуги аорты, врождённые пороки сердца, КТ-ангиография, Z-показатель, аортальное отношение, детская сердечно-сосудистая визуализация, измерения аорты, диагностическая согласованность.