

## Diagnostic Ability of Cardiovascular Computed Tomography (CCT) and Echocardiography for Aortic Anomalies in Paediatric Patients

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### Abstract

Aortic abnormalities are uncommon but potentially life-threatening disorders that require proper diagnosis and treatment. Aortic abnormalities in infants are predicted to occur in 2-6/10,000 live births, with aortic coarctation being the most prevalent one. Radiological imaging is essential in the identification and treatment of juvenile aortic abnormalities. Cardiovascular computed tomography (CCT) and echocardiography (Echo) are two imaging modalities that are frequently utilized to diagnose aortic abnormalities in pediatric patients. Both procedures have advantages and disadvantages, and they are typically used in conjunction to provide a complete examination of aortic anatomy and function. In this paper, we will discuss the diagnostic capability of Cardiovascular Computed Tomography (CCT) and Echocardiography for aortic abnormalities in paediatric patients, with a focus on their surgical implications.

### Introduction

Congenital aortic abnormalities are an uncommon group of cardiovascular illnesses caused by abnormal branchial arches during development. Aortic abnormalities in infants are predicted to occur in 2-6/10,000 live births, with aortic coarctation being the most prevalent one. Echo and CCT are complementary imaging techniques for the detection of aortic abnormalities. Echo can clearly define intracardiac anomalies, but it cannot accurately assess extracardiac structures such as arch of aorta, distal pulmonary arteries, and related vessels due to weak acoustic shadowing. CCT is a non-invasive approach that allows for extensive anatomical coverage and excellent spatial resolution for distal vascular structures, as well as the detection of concomitant airways or oesophageal compressions [1,2]. CCT enables accurate assessment of dimensions, site, and extension of aortic anomalies for surgical planning in terms of placement of surgical clamping and the extent of resection [3].

The literature search indicated a paucity of studies in the Indian population in terms of detailed evaluation (involvement, site, and extension) of aortic anomalies.

In arch repair surgeries, Information gathered from any investigative tool must provide not only the diagnosis but also surgical insights about the safety and site of clamp applications.

Additionally, CT Brain angiography was also done concurrently in our study to evaluate the completeness of Circle of Willis for prognostication and surgical planning in arch repairs.

## Aims and Objectives

- To investigate and compare the diagnostic findings of CCT and Echo for the comprehensive detection of aortic abnormalities in pediatric patients.
- To assess agreement between CCT, echo, and intra-operative findings.

## Study Methodology

This study was carried out at a tertiary level hospital. We enrolled retrospective paediatric cases from Jan 2011 to October 2019 and prospective cases from October 2019 to June 2022 with suspected aortic anomalies. A total of 110 paediatric patients who presented to the Paediatric Cardiac Sciences department and underwent Echo and CCT, were included. The results were correlated with surgical findings which were considered confirmatory. The study excluded patients who were contraindicated to contrast media and lacked either Echo/CCT/intraoperative data. CCT was performed on Philips Ingenuity 128-slice scanner. Those patients which required sedation, were subjected to pre-anaesthetic check-up and sedated. Those who were on ventilator already, were taken on ventilator only. The Patients were scanned in supine position and scout films taken from the level of vertex to orbitomeatal line for CT brain and from thoracic inlet to first lumbar vertebrae for CT thorax. In this scan, non-contrast pictures were collected first, and then contrast was injected at 2 ml/kg, at a rate of 2-3ml/sec, followed by 10-20 ml saline flush using a pressure injector. CT brain angiography was also conducted in all cases at the same time to ensure the Circle of Willis was complete. Paediatric Cardiologists performed echo on patients using a Phillips IE 33 X- matrix ultrasound system with either S7-2, S5-1, or S12-4 probes, depending on the child's size and the anomaly to be examined. The aorta was separated into three primary divisions: ascending aorta, arch and descending aorta. We interrogated a total of 1540 points of observations in all three divisions of the Aorta.

The following points were assessed on both modalities and compared with the surgical findings:

### Points of Observation in different segments of thoracic Aorta:

#### A. Ascending Aorta

1. Root of aorta (Dilated vs Hypoplastic).
2. Sino-tubular junction effacement.
3. Tubular Ascending Aorta (Dilated vs Hypoplastic).

#### B. Arch

1. Involvement (Hypoplastic vs Dilated).
2. Location of involvement (Proximal vs Distal vs Total).
3. Brachiocephalic branch origin pattern (Normal vs Mirror imaged vs Aberrant).
4. Ease of clamping of the arch between 1st and 2nd branches.
5. Ease of clamping of the arch between 2nd and 3rd branches.
6. Type of Interruption (type A/B/C).
7. Vascular ring - Yes/No.

### C. Descending Aorta

1. Involvement (Discrete vs Diffuse).
2. Ductus Arteriosus (Patent vs Closed).
3. Location of CoA (Pre or Juxtaductal vs Postductal).
4. Isthmus (Hypoplastic vs Normal).

Cardiac structures were categorized into normal, hypoplastic, or dilated using Z scores (normal as -2 to +2) [4].

Discrete coarctation is described as focal constriction of the descending aorta. A diffuse coarctation is one that involves a relatively long segment of the aortic arch [5].

During surgical repair, at least 3 mm of physical space in the selected site of the arch between any two brachiocephalic vessels is required to apply a vascular occlusion clamp (ease of clamping). This ensures that the blood flow through the origin of the vessel proximal to the clamp is not compromised and simultaneously also leaves enough aortic tissue distal to the clamp for taking sutures safely. (Figure 1) Circle of Willis was taken incomplete when there is an anatomical discontinuity between any vessel forming the circle. It could be considered normal anatomical variant of the circle of Willis [6].



**Figure 1:** MDCT coronal reconstruction showing (a) presence of clamp ability (b) absence of clamp ability. Criteria for clamp ability was defined as minimum 3mm distance between branches of arch of aorta. **Figure (1a):** Shows presence of clamp ability whereas. **Figure (1b):** Shows absence of clamp ability.

### Statistical Analysis

The statistical parameters such as agreement (kappa characteristics) and concordance on both modalities were correlated with intraoperative findings for each point of observation. Quantitative data was presented using Mean and Standard Deviation. The study groups were compared using an unpaired t-test based on the results of the normality test. The qualitative data was presented using frequency and percentage tables. The Fisher test, student's t-test, and Chi-squared test were used to analyze the association between the research groups. 'P' values less than 0.05 were considered significant.

- Kappa < 0: No agreement
- Kappa between 0.00 and 0.20: Slight agreement
- Kappa between 0.21 and 0.40: Fair agreement
- Kappa between 0.41 and 0.60: Moderate agreement
- Kappa between 0.61 and 0.80: Substantial agreement
- Kappa between 0.81 and 1.00: Almost perfect agreement.

## Results

The demographic data of our 110 patients is tabulated in table 1.

<b>Table1:</b> Demographic Table (n=110 patients).	
<b>Age</b>	Day 1 – 18 years (mean age- 15 months)
	>90% (neonates and infants)
<b>Gender</b>	
Male	70(63.6%)
Female	40(36.3%)
<b>Clinical features</b>	
Dyspnea	86(78.3%)
Cyanosis	51(46.8%)
Low CO	49(45.6%)
Respiratory Infection	23(21.3%)

The evaluated parameters of Ascending Aorta on Echo, CCT, and surgery were tabulated in Table 2. Open heart surgery was performed for ascending aortic anomalies in 20/110 patients (18.1%) while in the rest of the patients, ascending aorta could not be evaluated as these patients had a closed heart surgery hence, only 20 patients were taken for evaluation for ascending aorta. A comparison chart of concordance and agreement(kappa) of both modalities with respect to surgical findings in the ascending aorta was depicted in Table 3.

<b>Table 2:</b> Evaluated parameters of Ascending Aorta on Echo, CCT and surgery(n=20).				
		<b>Echocardiography</b>	<b>CCT</b>	<b>Operative notes</b>
<b>Ascending Aorta</b>	<b>1. Aortic root</b>	Number (%)	Number (%)	Number (%)
	Normal	18(90%)	16(80%)	15(75%)
	Hypoplastic	2(10%)	4(20%)	5(25%)
	Dilated	0(0.00%)	0(0.00%)	0(0.00%)
	<b>2. Sinu-tubular junct. Effacement</b>			
	No	20(100%)	19(95%)	19(95%)
	Yes	0(0.00%)	1(5%)	1(5%)
	<b>3. Tubular AA</b>			
	Normal	18(90%)	18(90%)	17(85%)
	Hypoplastic	2(10%)	2(10%)	3(15%)
	Dilated	0(0.00%)	0(0.00%)	0(0.00%)

<b>Table 3:</b> Representation of comparison of overall concordance and agreement(kappa) between Echo and CCT in Ascending Aorta (n=20).						
	Parameters	Percent of concordance of findings between echo and operative notes(n=20)	Kappa coefficient between echo and operative notes	Percent of concordance of findings between CCT and operative notes(n=20)	Kappa coefficient between CCT and operative notes	P Value
Ascending Aorta	Aortic Root	17(85%)	0.560	19(95%)	0.714	0.621
	Sinu-tubular junc. Effacement	19(95%)	0.000	20(100%)	1.00	1.000
	Tubular AA	19(95%)	0.796	19(95%)	0.796	1.000

The evaluated parameters of Arch of Aorta on echo, CCT and surgery were tabulated in table 4.

<b>Table 4:</b> Evaluated parameters of Arch of aorta on Echo, CCT and surgery(n=110).				
		Echocardiography	CCT	Operative notes
Arch of Aorta	<b>1. Involvement</b>	<b>Number (%)</b>	<b>Number (%)</b>	<b>Number (%)</b>
	Normal	82(74.50%)	74(67.30%)	75(68.20%)
	Hypoplastic	28(25.50%)	36(32.70%)	35(31.80%)
	Dilated	0(0.00%)	0(0.00%)	0(0.00%)
	<b>2. Location</b>			
	Normal	82(74.50%)	74(67.30%)	75(68.20%)
	Proximal	1(0.90%)	1(0.90%)	0(0.00%)
	Distal	26(23.60%)	31(28.20%)	29(26.40%)
	Total	1(0.90%)	4(3.60%)	6(5.50%)
	<b>3. Brachiocephalic br. Org.</b>			
	Normal	84(76.40%)	71(64.50%)	72(65.50%)
	Mirror imaged	10(9.10%)	15(13.60%)	14(12.70%)
	Aberrant	16(14.50%)	24(21.80%)	24(21.80%)
	<b>4. Ease of clamping (1<sup>st</sup> and 2<sup>nd</sup>)</b>			
	No	7(6.40%)	16(14.50%)	13(11.80%)
	Yes	103(93.60%)	94(85.50%)	97(88.20%)
	<b>5. Ease of clamping (2<sup>nd</sup> and 3<sup>rd</sup>)</b>			
	No	10(9.10%)	12(10.90%)	12(10.90%)
	Yes	100(90.10%)	98(89.10%)	98(89.10%)
	<b>6. Interruption</b>			
	No	95(86.40%)	95(86.40%)	95(86.40%)
	Yes (type A)	9(8.20%)	9(8.20%)	9(8.20%)
	Yes (type B)	5(4.50%)	5(4.50%)	5(4.50%)
	Yes (type C)	1(0.90%)	1(0.90%)	1(0.90%)
	<b>7. Vascular ring</b>			
	Yes	5(4.50%)	5(4.50%)	6(5.50%)
	No	105(95.50%)	105(95.50%)	104(94.50%)

Comparison of concordance and agreement (kappa) between both modalities for Arch of Aorta was comprised in table 5. The evaluated parameters of descending aorta on Echo, CCT and surgery were tabulated in table 6. Comparison of concordance and agreement(kappa) between both modalities for descending aorta was comprised in table 7.

Post-operative echocardiographic evaluations on the first day did not reveal any residual defects or significant gradients in the ascending aorta or arch and none of the patient's required re-exploration in the immediate post operative period.

<b>Table 5:</b> Representation of comparison of overall concordance and agreement(kappa) between Echo and CCT in Arch of Aorta(n=110).						
	Parameters	Percent of concordance of findings between echo and operative notes(n=110)	Kappa coefficient between echo and operative notes	Percent of concordance of findings between CCT and operative notes(n=110)	Kappa coefficient between CCT and operative notes	P value
<b>Arch of Aorta</b>	<b>Involvement</b>	99(90%)	0.757	102(92.7%)	0.865	0.828
	<b>Location</b>	93(84.50%)	0.640	93(84.50%)	0.746	1.000
	<b>Brachiocephalic br. Origin</b>	95(86.30%)	0.702	107(97.20%)	0.947	0.005*
	<b>Ease of clampability (1<sup>st</sup> and 2<sup>nd</sup>)</b>	99(90%)	0.428	104(94.50%)	0.769	0.312
	<b>Ease of clampability 2<sup>nd</sup> and 3<sup>rd</sup>)</b>	103(93.60%)	0.661	107(97.20%)	0.865	0.332
	<b>Interruption</b>	110(100%)	1.00	110(100%)	1.00	1.000
	<b>Vascular Ring</b>	96(87.2%)	0.564	105(95.40%)	0.854	0.005*

<b>Table 6:</b> Evaluated parameters of Descending Aorta on Echo, CCT and surgery(n=110).				
		Echocardiography	CCT	Operative notes
<b>Descending Aorta</b>	<b>1. Involvement</b>	<b>Number (%)</b>	<b>Number (%)</b>	<b>Number (%)</b>
	Normal	64(58.20%)	73(66.40%)	69(62.70%)
	Discrete	36(32.70%)	32(29.10%)	30(27.30%)
	Diffuse	10(9.10%)	5(4.50%)	11(10.00%)
	<b>2. Ductus Arteriosus</b>			
	Patent	80(72.70%)	89(80.90%)	91(82.70%)
	Closed	30(27.30%)	21(19.10%)	19(17.30%)
	<b>3. Location of CoA</b>			
	Normal	64(58.20%)	67(60.90%)	68(61.80%)
	Juxtaductal	44(40.00%)	41(37.20%)	40(36.40%)
	Postductal	2(1.80%)	2(1.80%)	2(1.80%)
	<b>4. Isthmus</b>			
	Normal	70(63.60%)	67(60.90%)	67(60.90%)
	Hypoplastic	35(31.80%)	32(29.10%)	35(31.80%)
	Coarcted	5(4.50%)	11(10.00%)	8(7.30%)
	Dilated	0(0.00%)	0(0.00%)	0(0.00%)

<b>Table 7:</b> Representation of comparison of overall concordance and agreement(kappa) between Echo and CCT in descending aorta (n=110).						
	Parameters	Percent of concordance of findings between echo and operative notes(n=110)	Kappa coefficient between echo and operative notes	Percent of concordance of findings between CCT and operative notes(n=110)	Kappa coefficient between CCT and operative notes	P value
<b>Descending Aorta</b>	<b>Involvement</b>	96(87.2%)	0.763	102(92.7%)	0.854	0.261
	<b>Ductus Arteriosus</b>	95(86.30%)	0.612	104(94.5%)	0.817	0.066
	<b>Location of CoA</b>	106(96.30%)	0.926	109(99.1%)	0.981	0.369
	<b>Isthmus</b>	92(83.60%)	0.678	98(89.1%)	0.794	0.231

## Discussion

In the current study, the most common anomaly was coarctation of the aorta (42/38.1% cases) followed by hypoplasia of the arch (35/31.8% cases). Coarctation of the aorta was discrete and juxtaductal in a majority of patients. Soleimantabar H et al [7] also conducted a retrospective evaluation of comparative accuracies of both modalities in aortic anomalies found coarctation to be most common aortic arch anomaly (19.7%). The following observations were noted:

### Ascending aorta:

The agreement for the diagnosis of ascending aortic hypoplasia was similar between both modalities with intraoperative findings however, for aortic root pathologies, the agreement was found slightly higher for CCT as compared with echo. This finding was dissimilar with Aiyin li et al [8] where 100% agreement was found for both modalities with intra-operative finding and we found more concordance and agreement for MDCT (95%) than Echo (85%) for aortic root pathologies, but our data was very limited to conclude affirmatively.

### Arch of Aorta:

Both agreement and concordance were found more accurate with CCT than Echo for anomalies/parameters of the arch of the aorta like arch hypoplasia and its location, brachiocephalic branch origin anomalies, vascular rings, and surgical clamp abilities (table 4). Statistically significant results (<0.05) between two modalities are seen for brachiocephalic branch origin anomaly and vascular ring parameters. S.M. Shehata et al [9] also proposed that MDCT could supplement Echo in presenting the aberrant arch structure and branching patterns required for surgical planning.

CCT also provided details about airway compressions, a bony framework of the chest, distal pulmonary vessels, any associated lung pathology etc, which can't be assessed on Echo. A combination of 3D reformatting and volume rendering can be used to determine the precise coverage of vascular ring and its external compression effects on the trachea, and aid in surgical planning.

Long et al [10] and Schulman [11] found that MDCT can better demonstrate anatomical connections and coverage between the trachea and vascular systems which help in assessing vascular rings.

Clamp ability between the branches of the arch of the aorta was evaluated in our study, which is an important parameter for surgical planning and approach.

For the surgical correction of the arch, a few important findings need to be evaluated preoperatively by Echo or CCT.

**1. Brachiocephalic branching pattern with emphasis on the single or dual origin of carotid arteries (normal branching v/s bovine v/s ovine branching).** It is imperative to perfuse at least one carotid while the repair is being undertaken. This implies that even if one carotid cannot be safely perfused, the surgical approach mandatorily shifts from thoracotomy (lateral) to mid-sternotomy (midline).



2. During lateral approach where brain perfusion is maintained by at least one patent carotid and ipsilateral subclavian artery, it is assumed that the circle of Willis is complete and the anterior circulation will provide for the lack of antegrade blood flow from the posterior circulation. Similarly, another aspect could be the right and left cross-circulation provided by an intact circle of Willis. In our study, 25.4% (28 patients) had an incomplete circle of Willis on CT. This is our theoretical assumption based on the anatomy of the circle of Willis being evaluated on CT to rule out all possibilities of vascular abnormalities such as the absence or stenosis of main vessels of brain circulation such as vertebral arteries, ICAs, or communicating vessels, in order to improve future poor neurological outcomes during arch surgeries or post-surgery by placing clamps between the arch vessels. Although in our study, we could not find any patient for which we changed any approach.

Safety profile based on the clamping ability of the aorta can be divided into 3 possible scenarios: -

1. Both carotids are perfused during surgery – the safest scenario and lateral thoracotomy can be used.
2. Only One carotid perfused (clamp between 1st and 2nd branch)- safety depends on careful analysis of the circle of Willis(complete/incomplete) and presence/absence of aberrant subclavian artery.
3. Proximal arch pathology and no possibility of carotid perfusion- midline approach has to be used.

### Descending Aorta:

CCT improved the accuracy of diagnosing aortic coarctation by demonstrating involvement (discrete/diffuse), location, and type of coarctation, as well as accompanying collateral development. Our results are similar to S.M. Shehata et al [9] who showed good agreement of MDCT ( $k = 0.870$ ) in detecting coarctation. Soleimantabar H et al [7] also found perfect agreement between the two modalities for detecting coarctation of the aorta (Kappa coefficient: 0.93%).

In our study, it was found that CCT can better demonstrate patency of ductus arteriosus as compared to echo with a perfect agreement (Kappa coefficient -0.817) with intra-operative findings. Although echocardiography is the preferred modality for diagnosing patent ductus arteriosus, CCT can better assess ductal morphology, size, and calcification. Zhao Q et al [12] also discovered that a dual source CT scan (DSCT) improved the imaging of extra-cardiac vascular abnormalities, including PDA.

It was found in our study that isthmus hypoplasia was slightly better identified by CCT with more accurate and concordant results (89.1%) than echo (83.60%). We could not find any comparative study of echo versus CCT for the assessment of isthmus pathologies in the literature.

Overall agreement coefficient (kappa) was found more with CCT than echo with respect to operative notes however, statistically significant ( $p < 0.05$ ) comparative results were found in terms of brachiocephalic branch origin patterns and vascular ring pathologies. Sensitivity= 85.7%, Specificity= 89.5%, PPV= 97.5%, NPV=56.7%, Diagnostic Accuracy= 86.4% were observed b/w echo and surgery (table 8). Sensitivity= 97.8%, Specificity= 81%, Positive predictive value=95.6%, Negative predictive value= 89.5%, Diagnostic accuracy= 94.5% were observed b/w CCT and surgery (table 9).

Table 8: Sensitivity of transthoracic echo with respect to operative notes.			
Echo/ Operative notes	Positive	Negative	Total
Positive	78	2	80
Negative	13	17	30
Total	91	19	110
Measure of Agreement	Kappa Value	0.612	P value= 0.00

Table 9: Sensitivity of CCT with respect to operative notes.			
CCT/ Operative notes	Positive	Negative	Total
Positive	87	4	91
Negative	2	17	19
Total	89	21	110
Measure of Agreement	Kappa Value	0.817	P value= 0.00



Limitations of our study include that it was a single-centre, single observer, hospital-based study. The sample size was small in terms of individual anomalies/parameters. There was a bias between the measurements of arch taken on both imaging modalities, which could not be eliminated. We had our hypothesis while planning the study about incomplete circle of willis and its implications. However, we did not have any patient where we had incomplete circle of willis with aberrant arch branching where clamping could not be done so, we may suggest assessment at a larger scale for this aspect.

## Conclusion

We conclude that CCT can supplement Echo in detailed evaluation of extracardiac anomalies for better surgical planning. The focussed approach should be there on each parameter including clamp space between arch branches, which may have implications. Evaluation of the circle of Willis in addition may have advantages and its impact on neurological outcomes needs to be evaluated on a larger scale. A comprehensive pre-operative analysis of the patients by Echo and CCT can help evaluate the therapeutic options, plan approach and extent of surgery.

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