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Diagnosis of Congenital Coarctation of the Aorta and Accompany Malformations in Infants by Multi-Detector Computed Tomography Angiography and Transthoracic Echocardiography: A Chinese Clinical Study

Authors' Contribution:
Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Background: The purpose of this study was to evaluate the utility of multi-detector computed tomography (MDCT) angiography and transthoracic echocardiography (TTE) in the diagnosis of congenital coarctation of the aorta (CoA) and accompanying malformations in infants.

Material/Methods: From January 2012 and December 2015, we enrolled 68 infants with clinically suspected CoA who underwent MDCT angiography and TTE in our hospital. Surgical correction was conducted to confirm the diagnostic accuracy of both examinations in all patients.

Results: In this study, the diagnosis of CoA was confirmed infants by surgical results in 55 of 68 infants. The diagnostic accuracy, sensitivity, and specificity of MDCT angiography were 95.6%, 96.4%, and 92.3%, respectively. The diagnostic accuracy, sensitivity, and specificity of TTE were 88.2%, 90.9%, and 76.9%, respectively. There was no significant difference in diagnostic accuracy, sensitivity, and specificity between MDCT angiography and TTE (χ^2=2.473, p>0.05, χ^2=1.373, p>0.05 and χ^2=1.182, p>0.05, respectively). In the diagnosis of concomitant cardiac abnormalities with CoA, the 2 methods also play different roles.

Conclusions: MDCT angiography and TTE play different roles in the diagnosis of CoA and accompany malformations. MDCT angiography in the diagnosis of the extra-cardiac vascular malformations is better than TTE, and TTE is superior to MDCT angiography in diagnosing intracardiac malformation. Combined MDCT angiography and TTE is a relatively valuable, reliable, and noninvasive method in the diagnosis of CoA and accompany malformations in infants.

MeSH Keywords: Aortic Coarctation • Echocardiography, Doppler • Multidetector Computed Tomography

Abbreviations: **CoA** – coarctation of the aorta; **MDCT** – multi-detector computed tomography; **TTE** – transthoracic echocardiography; **MPR** – multiplanar reformatting; **MIP** – maximum intensity projection; **VR** – volume rendering

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Background

Coarctation of the aorta (CoA) is a partial- or long-segment narrowing of the aorta, usually at the aortic isthmus, between the ductus arteriosus and the left subclavian artery. CoA was first described in 1760 by Morgagni in a dissected cadaver [1]. The incidence of CoA is 5% to 7% of cases with congenital heart disease and 0.03% to 0.04% of live births [2]. CoA is often accompanied by other congenital heart malformations, including bicuspid aortic valve (reported in about 40% of patients), ventricular septal defect, patent ductus arteriosus, mitral valve malformation, atrial septal defect, persistent left superior vena cava, right aortic arch, supravalvular stenosis of pulmonary artery, transposition of the great vessels, and anomalous unilateral pulmonary vein [3]. Therefore, early and accurate diagnosis is of great significance to the treatment and prognosis of CoA and accompanying malformations in infants. The purpose of this study was to compare the value of multi-detector computed tomography (MDCT) angiography and transthoracic echocardiography (TTE) for the diagnosis of congenital CoA and accompanying malformations in infants in a Chinese cardiac center.

Material and Methods

Approval was obtained from the Institutional Review Board of the University of Fujian Medical University, China, for a retrospective review of infantile patients with clinically suspected CoA who received MDCT angiography and TTE. Additionally, written parental informed consent was signed by the parents of the patients.

We reviewed the charts of 68 consecutive infants with clinically suspected CoA who were admitted to our hospital between January 2012 and December 2015 and who were subjected to MDCT angiography and TTE. All the infants were symptomatic, which included shortness of breath, dyspnea, fever, cough, expectoration, feeding difficulties, and developmental delays. Standard demographic information was collected (Table 1). There were 25 females and 43 males. The patients were aged from 15 days to 6 months (mean ± standard deviation, 2.1±1.8 months). Their weights ranged from 3.5 to 6.5 kg (4.5±1.6 kg), all infants were weighed to enable calculation of the appropriate chloral hydrate dose. Chloral hydrate was administered orally or rectally to each infant at doses of 50 mg/kg for sedation by an anesthesiologist.

MDCT angiography examinations were performed with a DSCT scanner (Somatom Definition; Siemens, Germany). Patients were examined while supine, and we took images extending from the base of the neck to the diaphragm. We used retrospective ECG-gated cardiac CT scanning. A low-dose protocol was used to reduce the radiation dose: collimation 0.6 mm; pitch 0.27–0.65, depending on the heart rate; rotation time 83 ms; slice thickness 1 mm; reconstruction interval 0.75 mm, and tube voltage 80 kVp. In all babies, an iodinated contrast medium, iopromide at 350 mgI/ml (Schering Ultravist, Iopromide, Berlin, Germany) was injected. The volume of contrast medium was adjusted to the body weight: 2–2.5ml per kg. The rate of injection was 0.5–1.0ml/s, depending on body weight. The scanning delay was determined with a bolus tracking technique. All images were transferred to an external workstation (Leonardo; Siemens Medical Solutions, Forchheim, Germany). 3D image reconstruction included multiplanar reformatting (MPR), curved planar reformatting, maximum intensity projection (MIP), and volume rendering (VR). (Figures 1, 2). A radiologist with 10 years' experience evaluated the data sets for each of the cases [4].

Table 1. Clinical data of 68 consecutive infants with clinically suspected CoA in this study.

Item	
Sex (M: F)	25: 43
Age (months)	2.1±1.8
Weight (kg)	4.5±1.6
Symptom	
Shortness of breath, dyspnea	55
Fever	12
Cough, expectoration	44
Feeding difficulties, developmental delays	60

The transthoracic echocardiography apparatus was a GE Vivid7 with 2.0–5.0 MHz transducer. Patients were examined in supine and left lateral position. The apical 4-chamber view, left ventricular long axis view, suprasternal view, and large artery short axis view were used in scanning, with special attention to abnormal intracardiac structure, the aorta, and its interconnections. Suprasternal views were used to evaluate the aortic arch and the connecting structure to determine if there was CoA and to measure its scope and inner diameter. Color Doppler imaging was used to evaluate the blood flow and measure the differential pressure and maximum speed in the location of the CoA [5] (Figures 3, 4). All TTEs were performed by a sonographer with 20 years' experience.

Statistical analysis

Continuous data are presented as means ± standard deviations and ranges. The time consumption of the 2 methods was compared with the independent-samples *t* test. The diagnostic

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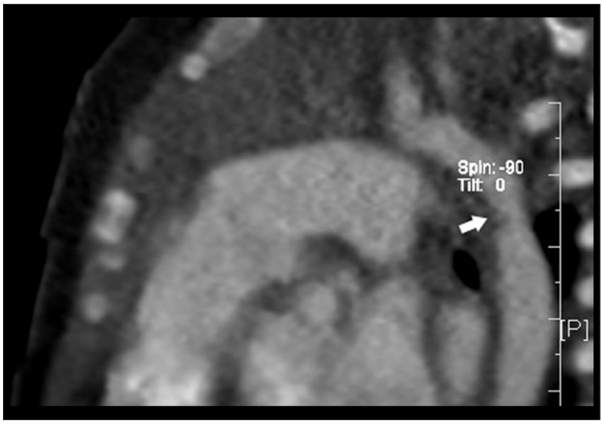

Figure 1. MDCT angiography showing the location of CoA in a 2-month-old (A) boy (arrow).

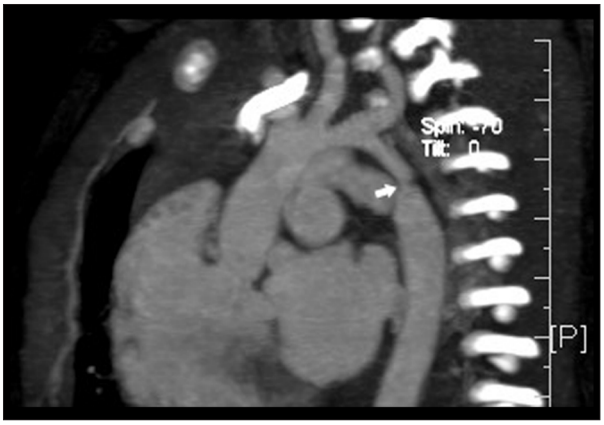

Figure 2. MDCT angiography showing the location of CoA in a 1-month-old (B) girl (arrow).

accuracy, sensitivity, and specificity of the 2 methods were analyzed by the chi-square test. A *P* value of <0.05 was defined as statistically significant.

Results

In this study, 55 infants had a diagnosis of CoA confirmed by surgical findings, and all 55 infants with CoA had the preductal type. The diagnostic accuracy of MDCT angiography and TTE was 95.6% and 88.2%, respectively. There was no significant statistical difference in diagnostic accuracy between MDCT angiography and TTE (χ^2=2.473, p>0.05). (Tables 2, 3). The diagnostic sensitivity and specificity of MDCT angiography for CoA were 96.4% (53/55) and 92.3% (12/13), respectively. The diagnostic sensitivity and specificity of TTE were 90.9% (50/55) and 76.9% (10/13), respectively. There was no statistically significant difference in diagnostic sensitivity and specificity between MDCT angiography and TTE (χ^2=1.373, p>0.05 and χ^2=1.182, p>0.05). The accuracy of the combined diagnostic protocol was 98.5%, which was significant better than

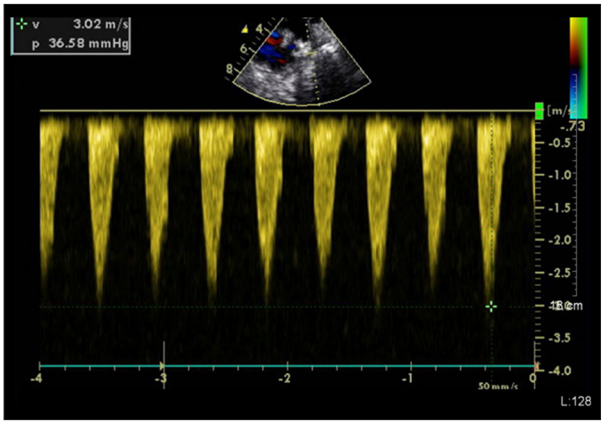

Figure 3. Transthoracic echocardiograms in suprasternal view in a 2-month-old (A) boy, showing high-speed flow in the CoA.

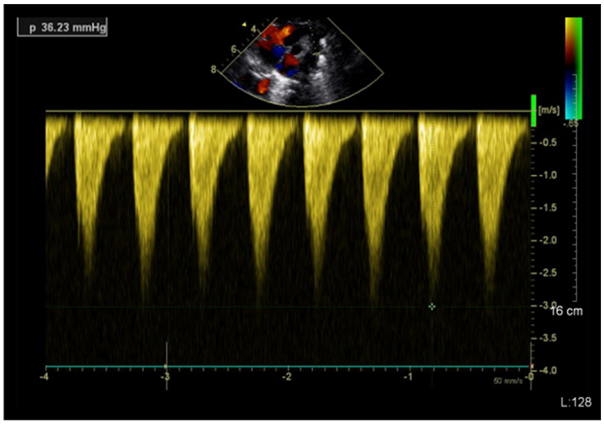

Figure 4. Transthoracic echocardiograms in suprasternal view in a 1-month-old (B) girl, showing high-speed flow in the CoA.

that of TTE (χ^2=5.83, p<0.05) and was not significantly different from MDCT angiography (χ^2=1.03, p>0.05).

In the diagnosis of concomitant cardiac abnormalities with CoA, these 2 methods play different roles. The 151 abnormal intracardiac structure malformations and the 31 extra-cardiac vascular malformations were confirmed by surgery (Table 4). In the 151 intracardiac structural abnormalities, 103 were confirmed and 48 were misdiagnosed by MDCT angiography, including mitral valve regurgitation in 8 cases, tricuspid valve regurgitation in 16 cases, bicuspid aortic valve in 19 cases, and patent foramen ovale or atrial septal defects in 5 cases. However, all the intracardiac structural abnormalities were detected by TTE. For intracardiac structural abnormalities, the diagnosis coincidence rates of these 2 methods were 68.2% (103/151) and 100% (151/151), which was a significant difference (p<0.001). All 31 extra-cardiac vascular malformations were confirmed by MDCT angiography. There were 11 lesions misdiagnosed by TTE, which included patent ductus arteriosus in 6 cases,

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Table 2. The comparison of the diagnosis of CoA at MDCT angiography with surgical results.

Item	Correct diagnosis	Misdiagnosis	Total
MTDCT angiography positive findings	53	1	54
MDCT angiography negative findings	2	12	14
Total	55	13	68

Table 3. The comparison of the diagnosis of CoA at TTE with surgical results.

Item	Correct diagnosis	Misdiagnosis	Total
TTE positive findings	50	3	53
TTE negative findings	5	10	15
Total	55	13	68

Table 4. Surgical findings in 55 patients with CoA.

Type	Counts
Coarctation with other cardiac malformations	55
Coarctation of solitary type	0
Coarctation of aorta-preductal type	55
Coarctation of aorta-postductal type	0
Associated intracardiac malformations	151
Ventricular septal defect	52
Patent foramen ovale or atrial septal defects	55
Anomalous pulmonary vein connection	1
Mitral valve regurgitation	8
Tricuspid valve regurgitation	16
Bicuspid aortic valve	19
Extra-cardiac vascular malformations	31
Aorto-pulmonary septal defect	3
Patent ductus arteriosus	23
Persistent left superior vena cava	5

persistent left superior vena cava in 3 cases, and aorto-pulmonary septal defect in 2 cases. For extra-cardiac vascular malformations, the diagnosis coincidence rates in these 2 methods were 100% (31/31) and 64.5% (20/31), which was a statistically significant difference ($p<0.001$).

None of the patients presented vascular complication or drug adverse effects when MDCT angiography and TTE were performed. The average time needed to finish MDCT angiography and TTE were 3.5–7 min (4.1±1.2 mins) and 25–45 min (32.1±6.2 min), respectively. The time needed to perform TTE was significantly longer than that needed to perform MDCT angiography ($p<0.05$). The cost, however, was significantly higher with MDCT angiography compared TTE (1250 *vs.* 285 RMB).

Discussion

Coarctation of the aorta is a relatively common congenital heart disease, which represents about 7% of total incidence. It is frequently associated with other cardiac abnormalities [6,7]. From the perspective of pathological physiology, CoA can be divided into 2 types: preductal and postductal. In this study, all patients had the preductal type. Clinical symptoms of CoA are heterogeneous, ranging from an asymptomatic patient with lower-limb hypotension and upper-limb hypertension to an ill newborn with repeated congestive heart failure. It normally presents severe symptoms during infancy, including shortness of breath, dyspnea, fever, cough, expectoration, breath-holding, and developmental delays. According some reports, CoA requires corrective surgery during the first year of life [8,9]. CoA is also frequently associated with other cardiac abnormalities, including mitral valve regurgitation, tricuspid valve regurgitation, bicuspid aortic valve, ventricular septal defect, patent foramen ovale or atrial septal defects, patent ductus arteriosus, left superior vena cava, and aorto-pulmonary septal defect [10]. In these cases, the infantile patients usually suffered from pulmonary infection and heart murmur before they were admitted to the hospital, and, to increase survival rate and improve the prognosis and quality of life, rapid and accurate diagnosis was necessary for these patients, which influenced further treatment planning.

There are a variety of inspection methods used for the diagnosis of CoA and accompanying malformations, including TTE, digital

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subtraction angiography, CT angiography, and MR angiography. Digital subtraction angiography is the criterion standard detection method, but it sometimes involves complications, including vascular injury and arterial thrombosis [11–13]. Moreover, it exposes both patient and staff to radiation. Recently, TTE and CT angiography have been shown to be less invasive and time-consuming than other methods, and have become commonly used in clinical practice.

TTE is a noninvasive, widely available, relatively low-cost, easily performed examinational that can be repeated frequently and it has been used in the preliminary diagnostic approach to CoA and accompanying malformations [14,15]. Its Doppler function can be used to identify the location and assess the severity of the coarctation and has the advantage of a noninvasive estimation of the pressure gradient across the narrowing [16]. Because of the poor acoustic window and the long distance between the transducer and the aorta isthmic region, it is sometimes difficult to obtain good visualization of the site of coarctation. In neonates and young infants, CoA may be missed or underestimated by TTE, especially in the presence of a large patent ductus arteriosus or severe pulmonary infection and dyspnea [17,18]. TTE cannot completely evaluate all the arch structure and the 3 branches of the aorta; however, it is one of the best tools for assessing hemodynamic changes in detecting the intracardiac structural abnormalities. It also provides much important information about other cardiovascular structures, hemodynamics, and cardiac function.

In the present study, the diagnostic accuracy, sensitivity, and specificity of TTE were 88.2%, 90.9%, and 76.9%, respectively. These parameters of TTE were lower than those of MDCT angiography, but the difference was not clinically significant. The diagnosis was missed in 5 patients, which shows that TTE has limitations in evaluating the length of the CoA and in visualizing collateral circulation, so MDCT angiography should be used in these patients, if necessary. Another factor, the diagnostic result of TTE, was strongly operator-dependent, depending on the extent of sonographer experience. Our results show that TTE has obvious advantages in the diagnosis of concomitant intracardiac abnormalities. Considering that CoA commonly co-occurs with other congenital heart defects [19,20], TTE should be the first choice for the preliminary diagnosis of CoA and accompanying malformations.

MDCT angiography has become an important imaging modality for the evaluation of aortic arch anomalies in infants. There are many advantages in using MDCT angiography for the diagnosis of CoA [21–24]. MDCT angiography has a very good spatial and 3D resolution and is less invasive. Using its 3D images, it can be highly accurate in assessing the overall structure of CoA, which can be analyzed and determined using many post-reconstruction techniques. The high image resolution is an important reason why CT angiography may be a helpful tool in the preoperative imaging evaluation of CoA. It can help surgeons to determine the location of the lesions and their surrounding structure, which can allow the surgeon to develop a suitable surgical plan [25,26]. The radiation dose needed in MDCT angiography has been reduced significantly, which makes it relatively safe for infants and young children. In addition, MDCT angiography is excellent at finding associated extra-cardiac vascular malformation anomalies, and it is the fastest of all noninvasive methods. However, it is not useful for visualizing the aortic gradient or other cardiac malformations.

In this study, the diagnostic accuracy, sensitivity, and specificity of MDCT angiography were 95.6%, 96.4%, and 92.3%, respectively, which is superior to those of TTE. The diagnosis was missed in 1 patient, who was diagnosed due to the different upper- and lower-limb blood pressures during the operation; therefore, it was necessary to monitor the onset of the upper- and lower-limb blood pressure of patients with clinically suspected CoA. In the present study, MDCT angiography was able to completely and clearly show extra-cardiac anatomy, and its diagnostic accuracy was significantly higher than that of TTE. Similar to results of other studies, we found that MDCT angiography has the advantages of showing the origin of the great vessels, the relationship between the spatial arrangement of vessels, and their connection to the heart, which can make up for the deficiencies of TTE.

There has been a recent increase in the number of studies comparing MDCT angiography with TTE in 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 DSCT angiography and TTE. Their results showed that the sensitivity, specificity, positive predictive value, negative predictive value, and overall diagnostic accuracy of DSCT in 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 DSCT was greater than that of TTE (62.5%; 50/80). On the contrary, for 38 cardiac anomalies, the sensitivity (78.9%, 30/38) of DSCT was lower than that of TTE (100%; 38/38) [27]. We obtained similar data in our study. A similar study by Nie et al. concluded that the diagnostic accuracy of TTE and DSCT angiography was 97.06% and 96.32%, respectively and they found no statistically significant difference in diagnostic accuracy between TTE and DSCT angiography (95.6% and 88.2%, respectively, $\chi^2=0$, $p>0.05$) [28]. We also obtained results with no statistically significant difference. Hu et al. reported the sensitivity of MDCT diagnosis for CoA was 100%, which was higher than that of color Doppler echocardiography (87.5%). They concluded that CTA with 3D reconstruction is a reliable, noninvasive method for the assessment of coarctation of the aorta [29]. Xin Chen et al. concluded that

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the diagnostic accuracy, sensitivity, and specificity of MDCT angiography for aortic arch anomalies were 99% (359/362), 100% (198/198), and 98% (161/164), respectively. The diagnostic accuracy, sensitivity, and specificity of TTE were 87% (315/362), 92% (182/198), and 81% (133/164, respectively) [21]. In summary, we arrived at the same conclusions as the studies cited above. Furthermore, we found there was better diagnostic accuracy in the protocol combining these 2 methods. Therefore, we recommend that MDCT angiography and TTE be jointly used in the diagnosis of CoA in infants.

MDCT scanning provides a complete inspection in a few seconds, which can reduce the use of sedative drugs and increase patient security. TTE examination, however, requires more time (over 30 min if the patient moves or is uncooperative). The cost of MDCT angiography is significantly higher than that of TTE. In our study, TTE also showed the advantage of lower cost than MDCT angiography in the diagnosis of CoA in infants. Comprehensive consideration of the diagnostic accuracy, sensitivity, and specificity, from the economic point of view in the current era, shows that TTE certainly should be an alternative method in light of escalating health care costs. Choice of an appropriate imaging modality for those infants with clinically suspected CoA must consider the safety, cost, availability, complications, and accompanying anomalies, as well as diagnostic accuracy, sensitivity, and specificity.

Like any retrospective study, ours included a possible bias associated with data collection and incomplete data from some patients. Although the number of infants in the study allowed us to produce statistically significant results, much larger numbers of patients must be evaluated to establish the results of diagnosis of CoA in infants by use of MDCT angiography and TTE. There may be more advanced methods that can replace these 2 methods and yield different results. Finally, this study was limited to a single institution, and other institutions may find different results.

Conclusions

In conclusion, MDCT angiography and TTE play different roles in diagnosing congenital CoA and can take the place of other examinations. The combination of MDCT angiography with TTE can improve the accuracy of diagnosis of CoA. This combined method is accurate in the diagnosis of CoA, and is a valuable guide in clinical decision-making.

Competing interests

The authors declare that they have no competing interests.

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