

The Role of Computed Tomography in The Diagnosis of Rare Congenital Heart Disease: Interrupted Aortic Arch

Yusen Feng,¹ Lijuan Wang,¹ Hui Liu,¹ Guifang Sun,¹ Bin Liu,¹ Pengcheng Ma,¹ Liqiong Wang,² Xuguang Zhang³

¹Department of Radiology, Kunming Yan'an Hospital; Key Laboratory of Cardiovascular Disease of Yunnan Province; Key Laboratory of Tumor Immunological Prevention and Treatment of Yunnan Province, Kunming City, China;

²Department of Pathology, Kunming Yan'an Hospital; Key Laboratory of Cardiovascular Disease of Yunnan Province; Key Laboratory of Tumor Immunological Prevention and Treatment of Yunnan Province, Kunming City, China;

³Department of Cardiac surgery, Kunming Yan'an Hospital; Key Laboratory of Cardiovascular Disease of Yunnan Province; Key Laboratory of Tumor Immunological Prevention and Treatment of Yunnan Province, Kunming City, China

ABSTRACT

Interrupted aortic arch (IAA) is a rare congenital anomaly of the aortic arch and an anatomical interruption of the lumen between the ascending and descending aorta. Computed tomography (CT) has become a reliable noninvasive diagnostic method for congenital IAA. The purpose of this study was to investigate the imaging features of IAA and improve the understanding and diagnosis of the disease. The imaging features and postoperative pathological data of 25 patients with IAA confirmed by dual-source computed tomography (DSCT) angiography were analyzed in this retrospective study.

Among the 25 patients with IAA, 15 were type A, seven were type B, 0 were types C and D, and two were type E. The diameter of the pulmonary artery trunk in type A was larger than that in type B ($P < 0.05$). However, there were no significant differences between types A and B along the ascending aorta diameter, descending aorta, ascending aorta/descending aorta ratio, left pulmonary artery main trunk diameter, right pulmonary artery main trunk diameter, left pulmonary artery trunk/pulmonary artery trunk ratio, right pulmonary artery trunk/pulmonary artery trunk ratio, and left pulmonary artery trunk/right pulmonary artery trunk ratio. The imaging findings of IAA have typical and specific signs, and the types of IAA are not comprehensive. One type of patient identification can be added: Patients who are dissected between the left common carotid artery and left subclavian artery opening, and the descending aorta is circulated by the chest collaterals. Patients with a wide pulmonary artery in the IAA are usually type A patients. Patients with IAA after surgical repair require lifetime follow up, mainly to monitor left ventricular outflow tract obstruction and recurrent aortic coarctation.

This study was approved by the Ethics Committee of Kunming Yan'an Hospital (Kunming City, Yunnan Province, China), and consent was waived because of the retrospective data collection.

INTRODUCTION

An interrupted aortic arch (IAA) is a rare congenital anomaly of the aortic arch and an anatomical interruption of the lumen between the ascending and descending aorta. Usually, it is associated with complex intracardiac malformations, such as patent ductus arteriosus (PDA), ventricular septal defect (VSD), or aortic valve lobular malformation. The disease was first reported by Raphael-Steidele in the late 18th century, and its proportion in infants with congenital heart disease was less than 1% [Alam 2009]. The incidence rate was 19 cases/1 million live births [Zhu 2018; Goudar 2016; Abdoli 2016; Benincasa 2015]. Failure to perform surgical intervention on the affected infant at an early stage will result in a mortality rate of 90% in the first year after birth [Sato 2011]. The majority of children with congenital IAA died of severe congestive heart failure, and hence, an appropriate diagnosis is critical for treatment. In recent years, with the development of multislice spiral technology, computed tomography (CT) has become a reliable noninvasive diagnostic method for congenital IAA. However, IAA is yet a rare occurrence. Radiologists have insufficient knowledge of the imaging manifestations of IAA, so it is easy to misdiagnose. Therefore, we collected the clinical, ultrasound, and CT imaging data of 25 patients with IAA and summarized and analyzed the imaging characteristics to improve the understanding of IAA, which in turn, would guide the decision-making of clinical treatment of the disease.

MATERIALS AND METHODS

General information: From April 2002 to August 2020, 25 patients, including 15 males and 10 females, with IAA confirmed by dual-source CT (DSCT) (definition flash, Siemens,

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Correspondence: Lijuan Wang, Telephone 0086- 15198808664 (e-mail: drwanglijuan@126.com).

Germany) angiography, in Kunming Yan'an Hospital were analyzed. The average age of the cohort was 9.01 years (range, 2 months–56 years). All patients underwent echocardiography before DSCT. Also, X-ray cardiography (collimator AL01C II Siemens, Germany) and ultrasonic were performed before the operation. Among them, 10 cases were treated by operation. All patients underwent DSCT reexamination within 3 months postoperatively. The patients were followed up by telephone after the operation. All patients are still alive. The flow diagram is shown in Figure 1. (Figure 1)

Inspection method: The Siemens flash 128-slice dual-source spiral CT scanner was used for plain and enhanced scans. Scanning parameters were as follows: tube voltage 120 kV, automatic Ma technology, detector combination of $2 \times 128 \times 0.6$ mm or $2 \times 64 \times 0.6$ mm, pitch 1.5, reconstruction layer thickness 1.0 mm, and spacing 0.6 mm. A volume of 60–80 mL contrast medium Iohexol Iodixanol Injection (H20103675, Jiangsu Hengrui Medicine Co., Ltd, China) (350 MG/mL) was injected through the median vein of the right elbow at a flow rate of 3–4 mL/s. At the end of the injection, 20–30 mL normal saline was injected into the tube at the same flow rate. The bolus tracking technique was used to select the aortic lumen as the target vessel for monitoring. When the enhancement threshold of CT value reached 120 Hounsfield unit (HU), the patients were asked to hold their breath and start scanning, ranging from the abdominal

aorta to the iliac artery. The images were sent to the workstation for multiplanar reorganization (MPR), curved projection reformation (CPR), maximum density imaging (MIP), and volume rendering (VR). The axial images were analyzed and observed.

Image processing: The images were transferred to Picture Archiving and Communication Systems (PACS) for MIP, including transverse, coronal, and sagittal images. The images of the ascending aorta, aortic arch, thoracic aorta, and abdominal aorta could be obtained by analyzing the sagittal section along the parallel aortic arch. The image needs MPR, CPR, and VR.

Image analysis: The film reading was analyzed by two attending physicians with more than 10 years of experience in cardiovascular imaging diagnosis. A consensus was established in the case of varied opinions. The location and shape of the aortic arch and the source of the severed descending aorta were analyzed. The diameters of ascending aorta, aortic arch, thoracic aorta, pulmonary artery trunk, left pulmonary artery trunk, and right pulmonary artery trunk was measured. The CT values of the area of interest of the ascending main artery and descending aorta in the arterial phase were measured by complete three-phase scanning.

Statistical analysis: SPSS 19.0 was used for the statistical analysis of the data. The normally distributed measurement data were expressed as standard deviation (sd). The independent sample t-test was used to compare the two groups of data. The count data were expressed by the number of cases (%), and the comparison between the groups was performed using the chi-square test. $P < 0.05$ indicated statistical significance.

RESULTS

Comparison of baseline data: A total of 25 IAA patients, including 15 males (60%) and 10 females (40%), were enrolled in this study. The average age of the cohort was 9.01 years (range, 2 months–56 years) and consisted of 17 patients with type A and eight patients with type B IAA. No significant difference was detected in the age ($F = 1.824$, $P = 0.19$), gender, and other general data between the type A and B groups ($P > 0.05$). (Table 1) Among them, 10 cases were treated by operation, and all were reexamined by DSCT within 3 months postoperatively. All patients survived.

In the cohort, 21 cases were diagnosed with IAA after physical examination, two cases were identified due to cough and fever, one case was found after dizziness, and one case was detected by echocardiography after cyanosis of lips. The systolic blood pressure (SBP) was 70–115 mmHg (1 mmHg=1.33

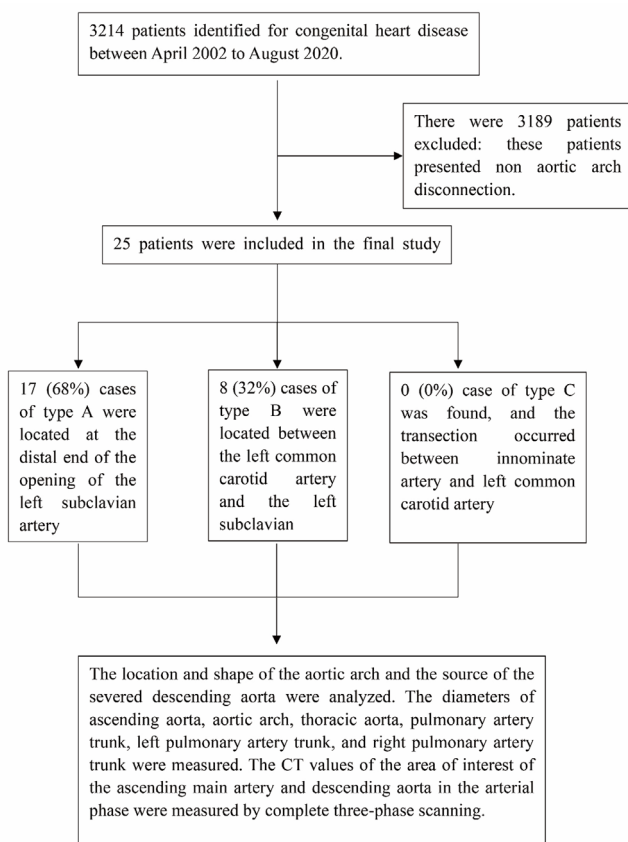


Figure 1. Flow diagram

Table 1. Baseline index of IAA patient

IAA patient types	Type A	Type B	P-value
Age	8.64±1.96	9.79±6.72	0.19
Sex (male/female)	10/7	5/3	0.861

kph), with an average of 88.45 mmHg, and the diastolic blood pressure (DBP) was 40–85 mmHg, with an average of 52.36 mmHg; the average SBP/DBP was 1.72.

Echocardiography results: Among the 25 patients with IAA, 20 patients showed aortic arch interruption by ultrasound; one case with ventricular septal defect, patent ductus arteriosus, atrial septal defect, and patent foramen ovale; four cases with ventricular septal defect, atrial septal defect, and patent ductus arteriosus; two cases with ventricular septal defect, patent ductus arteriosus, and patent foramen ovale; one case with ventricular septal defect and atrial septal defect; nine cases with ventricular septal defect and patent ductus arteriosus; one case with patent ductus arteriosus and patent foramen ovale; three cases with the ventricular septal defect; one case with the double outlet of the right ventricle; two cases with pulmonary aneurysm-like expansion; one case with persistent left superior vena cava; and two cases without other abnormalities.

Furthermore, among the 25 IAA patients, one case had bicuspid aortic valve malformation, one case had a tricuspid septal cleft, one case had aortic insufficiency and stenosis, two cases had aortic insufficiency, one case had aortic stenosis, 15 cases had mitral insufficiency, 20 cases had tricuspid insufficiency, one case had pulmonary valve stenosis and insufficiency, and one case had pulmonary insufficiency.

DSCT examination results: DSCT was used to diagnose 25 cases of IAA and the associated malformations. The development of the aortic arch, blood supply vessels of descending aorta, and other malformations can be shown on axial images. The anatomical structure of IAA could be displayed on the scanning images of the artery. There are three types, according to the different dissected parts of IAA [Oppenheimer-Dekker 1982]. Among them, IAA also is divided into six subtypes, according to the position of the right subclavian artery opening. The descending aorta is originated from the pulmonary artery. Type A with 15 (60%) cases is distal to the opening of the left subclavian artery, type B with five (20%) cases was severed between the left common carotid artery and the left subclavian artery, two (8%) cases of type B1 were located between the left common carotid artery and the left subclavian artery, and the right subclavian artery originated from the descending aorta. Type C was not detected in any case (0%), which was extremely rare; the type is located between the innominate artery and the left common carotid artery. However, three (12%) cases presented aortic circulation, which could not be classified, according to the above method. According to Wang et al. [Wang 2019], the source of blood supply in the descending aorta is related to cyanosis. Furthermore, IAA was divided into five types as follows (Figure 2): 15 (60%) cases were type A that were located at the distal end of the opening of the left subclavian artery; seven (28%) cases were type B, located between the left common carotid artery and left subclavian artery. (Figure 2) Also, there were 0 (0%) cases of type C, which was extremely rare, and the transaction occurs between the innominate artery and left common carotid artery. Furthermore, 0 (0%) cases of type D also was extremely rare; the severed part is located at the distal end of the opening of the left subclavian artery,

and the descending aorta is supplied by abdominal collateral branches, and two (8%) cases were type E, which also is extremely rare. The severed part is located at the distal end of the opening of the left subclavian artery, and the descending aorta is supplied by the thoracic collateral branch. One (4%) case does not belong to the above classification, the interruption was located between the left common carotid artery and the left subclavian artery, and the descending aorta was supplied by the thoracic lateral branch. (Figure 3) In this study, 17 (68%) cases of type A were located at the distal end of the opening of the left subclavian artery; eight (32%) cases of type B were located between the left common carotid artery and the left subclavian artery; and type C was found in 0 (0%) cases, with the transaction occurring between the innominate artery and left common carotid artery.

There is no direct blood flow communication between the left common carotid artery and left subclavian artery, which is connected by the residual fiber bundles. Then, the diameter of the ascending aorta was about 3.7 cm, which directly continued to the brachiocephalic trunk and the left common carotid artery. The inner diameter of the descending aorta is about 1.9cm, and it sends out the left subclavian artery. The inner diameter of the abdominal aorta is about 1.3cm, and the right subclavian artery and the brachiocephalic artery are tumor-like expansion, and the diameters are about 1.4 cm and 3.7 cm, respectively. Two large vessels (0.9 cm and 7 cm in diameter) from the right subclavian artery tortuously passed

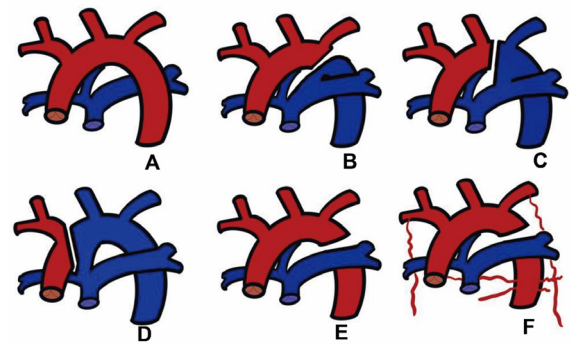


Figure 2. IAA classification diagram. (A) Schematic diagram of a normal aortic arch. (B) Schematic diagram of type A IAA. The interruption is located at the distal end of the left subclavian artery, and the descending aorta is supplied by the pulmonary artery. (C) Schematic diagram of type B IAA. The interrupted part is located between the left carotid common and left subclavian artery, and the descending aorta is supplied by the pulmonary artery. This type often is combined with the right subclavian artery and often vagus, and the descending aorta is supplied by the pulmonary artery. (D) Schematic diagram of type C IAA. The interrupted part is located between the innominate artery and the left common carotid artery, and the descending aorta is supplied by the pulmonary artery. (E) Schematic diagram of type D IAA. The severed part is located between the innominate artery and the left common carotid artery. The descending aorta is circulated for blood supply from abdominal collaterals. (F) Schematic diagram of type E IAA. The severed part is located between the innominate artery and the left common carotid artery. The descending aorta is circulated for blood supply from chest collaterals.

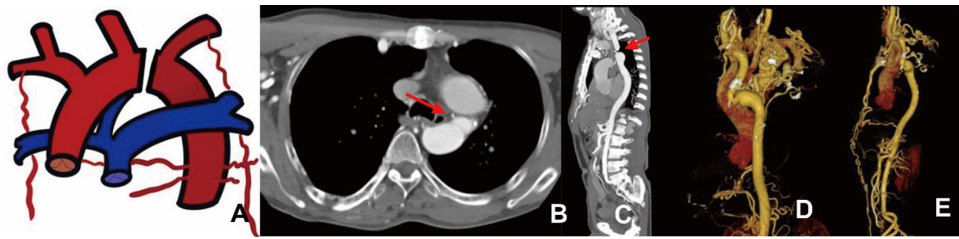


Figure 3. Special IAA classification diagram of a 56-year-old male patient. (A) Schematic diagram of the patient's dissected position and the source of the descending aorta. The dissected part is located between the left carotid artery and the left subclavian artery. The descending aorta is supplied by the systemic artery. (B) The transverse section image of CT vessel enhancement. The aortic arch and descending aorta are severed (pointed by the long red arrow). (C) The sagittal image of the CT vascular enhancement. The aortic arch and descending aorta are severed (pointed by the short red arrow). (D, E) Three-dimensional reconstruction images of blood vessels.

through the right cervical root. The left internal mammary artery was tortuous, thickened, and dilated to communicate with the left subclavian artery. The left internal mammary artery was tortuous and thickened, with tumor-like expansion. The left internal mammary artery was tortuous and dilated, and the local aneurysm was formed, with the maximum diameter of 19 mm; the tortuous and enlarged vascular mass at the level of diaphragm communicated with the celiac trunk. The tortuous and dilated superficial abdominal arteries communicated with the ipsilateral femoral artery; the tortuous and dilated vascular mass of the right axillary artery communicated with the right expanded intercostal artery (Figure 3D, 3E).

The average diameter of the main pulmonary artery in patients with type A was 3.27 cm, and in patients with type B, it was 2.81 cm. The average diameter of the main pulmonary artery in patients with type A was significantly larger than that in patients with type B ($P < 0.05$). The average diameter of the ascending aorta was 1.32 cm in type A patients and 1.41 cm in type B patients; the average diameter of descending aorta was 1.23 cm in patients with type A and 1.13 cm in patients with type B. The ratio of ascending aorta to descending aorta was 1.23 in patients with type A and 1.31 in patients with type B. The average diameter of the left main pulmonary artery was 1.40 and 1.26 cm in patients with type A and B, respectively. The diameter of the right main pulmonary artery was 1.48 cm in type A and 1.41 cm in type B. The ratio of the left main pulmonary artery diameter to pulmonary artery diameter was 0.45 in type A and type B, respectively, and the ratio of the right pulmonary artery diameter to the pulmonary artery diameter was 0.48 in type A and 0.50 in type B. Furthermore, the ratio of the left pulmonary artery diameter to the right pulmonary artery diameter was 0.94 in type A and 0.92 in type B.

In type A patients, the number of descending aorta sources (systemic circulation/pulmonary circulation) was 2/5, and that of type B was 1/7. The average CT value of the ascending aorta in type A was 414 HU, and that of type B was 289 HU. The average CT value of descending aorta in type A was 347.83 HU and that of type B was 394.75 HU. The CT value of ascending aorta/descending active pulse of type A was 1.17 and that of type B was 0.75. No significant difference was detected between the above data ($P > 0.05$). (Table 2)

Surgical findings: A total of 10 patients underwent

surgical treatment and DSCT reexamination within 3 months after the operation, including one case of type A and nine cases of type B. The average diameter of the ascending aorta was 1.28 cm before the operation and 1.44 cm after the operation. The average diameter of the descending aorta was 1.24 cm before the operation and 1.18 cm after the operation. The preoperative average diameter of ascending aorta/descending aorta ratio was 1.33, and the postoperative ratio was 1.26. The internal diameter of the main pulmonary artery was 3.30 cm before operation and 2.60 cm after operation. No significant difference was detected between the above values ($P > 0.05$). (Table 3) The postoperative diagnosis of type A and type B patients was consistent with that of DSCT angiography. Patients underwent deep hypothermic circulatory arrest under general anesthesia and selective cerebral perfusion for correction of aortic arch disconnection, and normal circulation postoperatively was restored.

In type A patients, the pericardium was cut off, and cardiopulmonary bypass was established. The left thoracic cavity was cut 4 cm away from the upper edge of the phrenic nerve. The 20 mm straight vessels were anastomosed to the 18 mm straight vessels with 5-0 Prolene lines, and the upper wall was clamped on the ascending aorta. The end-to-side anastomosis of Prolene line to the ascending aorta effectuated cooling, blocking the superior and inferior vena cava and the ascending aorta, making full flow bypass, and perfusing the aortic root (4°C high potassium cardioplegia solution). This makes the heart stop beating satisfied. We made the ligation of the arterial catheter, cut the pulmonary artery, and used 4-0 prolene double-ended needle with a gasket to close the arterial catheter, and then the pulmonary artery was formed. After rewarming and opening the ascending aorta, the heart automatically returned to beating, sinus rhythm. No adverse reactions occurred after protamine neutralization.

Patients with type B were heparinized to establish cardiopulmonary bypass (CPB) (via ascending aorta cannula and proximal descending aorta), cooling, blocking superior and inferior vena cava, and perfusing the aortic root (4°C high potassium cardioplegia solution). Consequently, cardiac arrest was satisfactory. A part of the main pulmonary artery was excised and sutured for autoplasty. The artificial blood vessel was constricted, and the other end of the anastomosed artificial blood vessel was pulled into the pericardial cavity

Table 2. DSCT data statistics of IAA patients (* $P < 0.05$)

IAA patient types	Type A	Type B	F-value	P-value
The inner diameter of ascending aorta (cm)	1.32	1.41	1.99	0.17
Inner diameter of descending aorta(cm)	1.23	1.13	0.54	0.47
The inner diameter of ascending aorta/descending aorta	1.25	1.31	0.95	0.76
Origin of descending aorta (systemic circulation/pulmonary circulation) (units)	2/15	1/7	$\chi^2 < 0.001$	0.96
Inner diameter main pulmonary artery (cm)	3.27	2.81	16.18	0.001*
The inner diameter of the left main pulmonary artery (cm)	1.4	1.26	2.24	0.15
The inner diameter of the right main pulmonary artery (cm)	1.48	1.41	0.02	0.89
Left main pulmonary artery inner diameter/main pulmonary artery inner diameter	0.45	0.45	0.10	0.76
Right main pulmonary artery inner diameter/main pulmonary artery inner diameter	0.48	0.5	0.08	0.78
Left main pulmonary artery inner diameter / right main pulmonary artery inner diameter	0.94	0.92	0.06	0.81
CT value of ascending aorta (HU)	414	289	1.60	0.24
CT value of descending aorta (HU)	347.83	394.75	5.11	0.05
CT value of ascending aorta/CT value of descending aorta	1.17	0.75	0.62	0.46

Table 3. Comparison of preoperative and postoperative data of IAA patients with DSCT. ($P > 0.05$)

	Preoperative data	Postoperative data	P-value
Ascending aorta inner diameter (cm)	1.28	1.44	0.38
Descending aorta inner diameter (cm)	1.24	1.18	0.48
Ascending aorta inner diameter/descending aorta inner diameter	1.33	1.26	0.59
Inner diameter of main pulmonary artery (cm)	3.30	2.60	0.91

through the left pulmonary hilum over the top of the pleura and the upper part of the pulmonary artery. An incision was made in the ascending aorta, and the other end of the artificial vessel was anastomosed with the ascending main artery with a 4-0 Prolene line for end-to-side anastomosis. After rewarming, adequate exhausting, and opening of the ascending aorta, the heart automatically resumes to sinus rhythm.

In this study, all 10 patients completed the surgery and survived.

DISCUSSION

IAA is a rare congenital cardiovascular malformation, accounting for 1% of the critical congenital heart diseases in children [Fyler 1980; Steidele 1778; Keane 1992]. The incidence rate of IAA is about 0.19/1000 live births, accounting for 5% of the cases of aortic arch obstruction [Alcantara 2020; Allen 2008]. The formation of IAA is related to chromosomal recombination and single gene abnormality [Martin 2003].

The formation of IAA is related to chromosome recombination and single gene abnormality [Martin 2003], which indicates an undeveloped, degenerated, and atrophic formation of the proximal segment or the fourth arch of the left dorsal aorta in the 6th to 7th week of an embryo. The

continuity of the lumen between the aortic arch and the descending aorta is interrupted. IAA rarely occurs in isolation. About 95% of the IAA patients also presented other complex congenital heart diseases, such as ventricular septal defect and atrial septal defect. About 8% of the patients were not accompanied by other heart diseases. In this study, only two cases (8.00%) were not accompanied by other heart diseases, and the IAA patients accompanied with ventricular septal defect and patent ductus arteriosus (nine cases, 36%) were the most, which was close to the above study [Martin 2003]. IAA was first described in 1778 and has a high mortality rate. Until the 1970s, with the emergence of prostaglandins and the improvement of surgical techniques, the complete repair of the neonatal period became normal [Steidele 1778; Keane 1992]. In this study, 10 patients completed surgical treatment, and all patients survived.

Clinical features of IAA: The anatomical abnormality of IAA is often associated with patent ductus arteriosus and ventricular septal defect also termed as IAA triad [Chen 2018]. The degree of collateral circulation in the chest, abdomen and lower limbs supplied by the descending aorta is the key to the patient's survival. When the patient's collateral circulation maintains basic lower limb function, the blood perfusion in the descending aorta mainly depends on the pulmonary artery, and lower limb hypoperfusion or hypotension may

occur, which can lead to electrolyte imbalance, renal insufficiency, multiple organ failure, or circulatory failure, and finally cause the patient to die in the neonatal period [Burbano-Vera 2018]. In this study, all adult patients had abundant collateral circulation. However, there were three patients in the study who were not supplied by the pulmonary artery but were only supplied by the chest or abdominal artery collateral circulation. One case was a 56-year-old adult, and two cases were children about 1 year in age. The clinical manifestations of adult IAA are diverse, including asymptomatic, headache, hypertension, and differences in blood pressure between upper and lower limbs [Peng 2019]. As the aortic arch is interrupted, the pressure of the ascending aorta is increased, and the blood pressure of the patient's upper limbs is significantly higher than that of the lower limbs. Therefore, if it is found that the blood pressure of the patient's upper and lower limbs is inconsistent, in addition to dissection and vascular stenosis, the possibility of IAA should also be considered.

Classification characteristics of IAA: IAA is characterized by the anatomical discontinuity between the ascending and descending aorta that can be complete discontinuity or connected by residual fiber band but interferes with the blood flow. The most commonly used method in clinical practice is described by Fournier et al. [Fournier 1959]. According to the different locations of disconnection, IAA is divided into the following three types: type A (accounting for 28%) is caused by the abnormal degeneration of the left fourth arch artery after the left subclavian artery rose to the normal position. In this study, 17 (68%) cases were type A, while type B (70%) is the most common type. The aortic arch breaks between the left common carotid artery and the left subclavian artery orifice, which is related to 22q11.2 deletion. This is caused by abnormal degeneration of the left fourth arch artery in the early development before the migration of the anterior part of the left subclavian artery. Type C (<5%) is rare; this involves the interruption of the aortic arch between the brachiocephalic trunk and opening of the left common carotid artery, caused by the abnormal degeneration of the left third and fourth arch arteries [Chan 2015].

Compared with the study by Fournier et al., there are more patients who presented type A in our study. This phenomenon could be attributed to the rare occurrence of IAA cases. Currently, there are no studies with a large dataset, and hence, there may be a deviation. Therefore, more studies are required to improve the epidemic-related data of this disease. Antje et al. [Oppenheimer-Dekker 1982] further divided IAA into six subtypes, according to different sites of interruption and the opening position of the right subclavian artery. However, the descending aorta originated from the pulmonary artery. In their study, 35% of IAA were type A and 6% were type A1. In our study, 15 (60%) cases of type A were severed at the distal end of the left subclavian artery orifice, and five (20%) cases of type B were located between the left common carotid artery and the left subclavian artery orifice. In the study by Arentje et al., 53% were type B and two cases were type B1 (8%). The transaction occurred between the left common carotid artery and the left subclavian artery opening, and the right subclavian artery originated from the

descending aorta, while in the study of Arentje et al., 6% of cases were type B1. In our study, rarely, type C was not clearly observed. Type C is located between the innominate artery and the left common carotid artery. Among them, three (12%) cases of the patients had an autologous circulation of the descending aorta, which could not be classified by the above classification method. Therefore, the author also classified, according to the research of Wang et al. [Wang 2019]. Because the source of blood supply in the descending aorta is related to cyanosis, Wang et al. classified IAA into five types, according to the analysis of disconnection position and blood supply of the descending aorta. In the present study, type A accounted for 60%, type B accounted for 28%, type C and type D accounted for 0%, and type E accounted for 8% of cases, which is extremely rare. The severed part is located at the distal end of the opening of the left subclavian artery, and the descending aorta is supplied by the thoracic collateral branches. Among them, one (4.00%) case did not belong to the above classification. The interruption of the one case occurred between the left common carotid artery and the left subclavian artery opening, and the descending aorta was supplied by the thoracic collateral branch circulation (Figure 3). Therefore, the author suggests adding a type G to the "two types and five types" (Figure 3A) classified based on cyanosis and non-cyanosis modified by Wang et al. [Wang 2019], which is located between the opening of the left common carotid artery and left subclavian artery, and the descending aorta is supplied by thoracic collateral branches. These adjusted classification criteria are in agreement with the conventional classification of congenital heart disease and are more comprehensive than those previously described.

Value of DSCT in the diagnosis of IAA: Currently, the noninvasive imaging methods for the diagnosis of IAA mainly include computed tomography angiography (CTA), echocardiography, and cardiac magnetic resonance (MR), which replace the invasive digital subtraction angiography (DSA) examination [Hanneman 2016]. Ultrasound is the preferred method for diagnosing congenital heart disease. It is characterized by non-radiation, real-time dynamics, and multiple levels. However, due to the high position of the aortic arch, the observation of this position is easily restricted. It takes a long time to identify patients with IAA by cardiac MR, which is not conducive to screening patients. A previous study [Chen 2018] reported that IAA is increased, as observed by CTA [Alam 2009]. Moreover, the anatomical continuity and separation between the aorta and the descending aorta were interrupted and separated. In some patients, strips of fiber could be observed. The ratio of ascending aorta diameter to descending main pulmonary artery diameter was abnormal, while the ascending aorta was widened, and the descending aorta was narrowed. The average value of our study was 1.27, indicating that the ascending aorta, descending aorta, and the ratio of ascending aorta/descending aorta of type A and type B patients were significantly different. Furthermore, in our study, the pulmonary artery diameter of type A patients is larger than that of type B patients. The authors speculate that it may be due to the pulmonary artery

supplying the descending aorta and the left subclavian artery perfusion in type B patients. Therefore, the pulmonary artery needs more pressure in type B patients than that in type A patients, who only have the descending aortic blood vessel, resulting in higher pulmonary artery pressure in type B patients, thus the type A patient's pulmonary artery has a wider inner diameter. There is no difference between type A and type B patients among the left and right pulmonary artery diameters, the ratio of left pulmonary artery diameter to main pulmonary artery diameter and the ratio of right pulmonary artery diameter to main pulmonary artery diameter, as well as the ratio of left pulmonary artery diameter to right pulmonary artery diameter, which might be caused by abundant collateral circulation shunt. Some patients also can see that it is supplied by the collateral circulation of tortuous and thickened arteries. For example, the descending aorta is supplied by the intercostal artery, celiac artery, left hepatic artery, internal iliac, and external arteries. There also are several cases in our study.

DSCT's multi-planar reconstruction, VR, and other three-dimensional reconstruction techniques can clearly determine the exact location of IAA and the relationship between adjacent blood vessels, and then determine the clinical classification and blood supply. In the study, it was found that sagittal imaging along the aortic arch is easier to show the location of the dissection. In addition, DSCT displays other congenital heart diseases and PDA shunt. DSCT combined with post-processing technology is used to design the operation path map for interventional therapy and evaluate the surgical effect [Goreczny 2017]. In the current study, 10 patients completed surgical treatment, and postoperative circulation recovered. Patients with IAA after repair require lifetime follow up, mainly to monitor the left ventricular outflow tract obstruction and recurrent coarctation of the aorta [Friedman 2018].

Treatment of IAA: Surgical reconstruction is the preferred treatment for IAA children. Immediate surgery is recommended if the hemodynamics are stable, and the postoperative effect is satisfactory, and although the reoperation rate is high, most children can survive for a prolonged period [Naimo 2017]. Whether IAA adults need surgical treatment is controversial, which needs to be combined with the patient's condition [Rodrigues 2017]. Because the adult patient survivors have abundant collateral circulation compensation, some patients can survive without surgery and can be treated conservatively.

Differential diagnosis of IAA: IAA should be differentiated from coarctation of the aorta (COA) and aortic arch thrombosis (AAT). COA refers to the local stenosis of the aortic cavity formed by the thickening of the inner wall of the aorta and the internal folding of the aortic wall tissue but has a complete vascular wall [Dijkema 2017]. NAAT is relatively rare, and most children cannot survive. It is also important to pay attention to other diseases of the patient, such as sepsis, other thrombotic diseases, polycythemia, etc., but the thrombus can be partially absorbed after anticoagulation therapy [Knadler 2019].

Limitations: The limitations of this study are as follows: (1) Since this is a rare disease studied with a small number

of cases, a large amount of data or meta-analysis is required. (2) The present study cohort does not encompass all types of cases; hence, additional samples are needed for research.

CONCLUSION

IAA is a rare congenital malformation. With typical imaging performance, combined with clinical and DSCT imaging and three-dimensional reconstruction, data can accurately diagnose IAA and determine the classification, collateral circulation, and cardiac malformations. The inner diameter of the pulmonary artery trunk in type A patients is wider than that in type B patients, which can provide a certain basis for their analysis. Other values, such as the inner diameter of the ascending aorta, descending aorta and pulmonary artery trunk, were not different between type A and type B patients. In terms of treatment, combined with image post-processing, technology can assist interventional therapy in evaluating surgical efficacy. Therefore, DSCT is of great value with diagnosing IAA.

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