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

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Abstract

Background: 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 To investigate the imaging features of IAA and improve the understanding and diagnosis of the disease.

Methods: 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.

Results: Among the 25 patients with IAA, 15 were type A, 7 were type B, 0 were type C and D, and 2 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 significantly different between type A and type B abomg 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; left pulmonary artery trunk/right pulmonary artery trunk ratio.

Conclusion: 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 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.

1 Introduction

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% [1]. The incidence rate was 19 cases/1 million live births [2−5]. 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 [6]. 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. Presently, the studies of IAA are mainly case reports. Radiologists have insufficient knowledge of the imaging manifestations of IAA and are easy to be misdiagnosed. 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, in which in turn, would guide the decision-making of clinical treatment of the disease.

2 Materials And Methods

2.1 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，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 IIsiemens，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. And all patients are still alive. The flow diagram was shown in Fig. 1.

2.2 Inspection method

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 × 128 × 0.6 mm or 2 × 64 × 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 IodixanolInjection (H20103675, Jiangsu Hengrui Medicine Co.,Ltd,china) (350 MGI/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), cerved projection reformation (CPR), maximum density imaging (MIP), and volume rendering (VR). The axial images were analyzed and observed.

2.3 Image processing

The images were transferred to Picture Archiving and Communication Systems (PACS) for MIP, including transverse, coronal, and sagittal images. The images of 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.

2.4 Image analysis

The film reading was analyzed by two attending physicians with more than10 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 were 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.

2.5 Statistical analysis

SPSS 19.0 was used for the statistical analysis of the data. The normally distributed measurement data were expressed as $\bar{x} \pm s$ 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.

3 Results

3.1 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 8 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 the patients had survived.

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

In the cohort, 21 cases were diagnosed with IAA after physical examination, 2 cases were identified due to cough and fever, 1 case was found after dizziness, and 1 case was detected by echocardiography after cyanosis of lips. The systolic blood pressure (SBP) was 70–115 mmHg (1 mmHg = 1.33 kpa), 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.

3.2 Echocardiography results

Among the 25 patients with IAA, 20 patients showed aortic arch interruption by ultrasound; 1 case with ventricular septal defect, patent ductus arteriosus, atrial septal defect, and patent foramen ovale; 4 cases with ventricular septal defect, atrial septal defect, and patent ductus arteriosus; 2 cases with ventricular septal defect, patent ductus arteriosus, and patent foramen ovale; 1 case with ventricular septal defect

and atrial septal defect; 9 cases with ventricular septal defect and patent ductus arteriosus; 1 case with patent ductus arteriosus and patent foramen ovale; 3 cases with ventricular septal defect; 1 case with double outlet of the right ventricle; 2 cases with pulmonary aneurysm-like expansion; 1 case with persistent left superior vena cava; 2 cases without other abnormalities.

Furthermore, among the 25 IAA patients, 1case had bicuspid aortic valve malformation, 1 case had a tricuspid septal cleft, 1 case had aortic insufficiency and stenosis, 2 cases had aortic insufficiency, 1 case had aortic stenosis, 15 cases had mitral insufficiency, 20 cases had tricuspid insufficiency, 1case had pulmonary valve stenosis and insufficiency, and 1 case had pulmonary insufficiency.

3.3 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 clearly displayed on the scanning images of the artery. There are 3 types according to the different dissected parts of IAA [8]. Among them, IAA is also divided into 6 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 5 (20%) cases was severed between the left common carotid artery and the left subclavian artery, 2 (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 was located between the innominate artery and the left common carotid artery. However, 3 (12%) cases presented aortic circulation, which could not be classified according to the above method. According to Wang et al. [9], the source of blood supply in the descending aorta is related to cyanosis. Furthermore, IAA was divided into five types as follows (Fig. 2): 15 (60%) cases were type A that were located at the distal end of the opening of the left subclavian artery; 7 (28%) cases were type B, located between the left common carotid artery and left subclavian artery. Also, there were 0 (0%) cases of type C, which was extremely rare, and the transection occurred between the innominate artery and left common carotid artery. Furthermore, 0 (0%) case of type D was also extremely rare, the severed part was located at the distal end of the opening of left subclavian artery, and the descending aorta was supplied by abdominal collateral branches, and 2 (8%) cases were type E, which is also 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. 1 (4%) case does not belong to the above classification, the interruption is located between the left common carotid artery and the left subclavian artery, and the descending aorta is supplied by the thoracic lateral branch (Fig. 3). In this study, 17 (68%) cases of type A were located at the distal end of the opening of the left subclavian artery; 8 (32%) cases of type B were located between the left common carotid artery and the left subclavian artery; type C was found in 0 (0%) cases, and the transection occurred between innominate artery and left common carotid artery.

The average diameter of the main pulmonary artery in patients with type A was 3.27cm, and that in patients with type B 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 and 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).

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

IAA Patient Types	Type A	TypeB	F value	P value
Inner diameter of ascending aorta (cm)	1.32	1.41	F = 1.99	0.17
Inner diameter of descending aorta(cm)	1.23	1.13	F = 0.54	0.47
Inner diameter of ascending aorta/descending aorta	1.25	1.31	F = 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	F = 16.18	0.001*
Inner diameter of left main pulmonary artery (cm)	1.4	1.26	F = 2.24	0.15
Inner diameter of right main pulmonary artery (cm)	1.48	1.41	F = 0.02	0.89
Left main pulmonary artery inner diameter / main pulmonary artery inner diameter	0.45	0.45	F = 0.10	0.76
Right main pulmonary artery inner diameter / main pulmonary artery inner diameter	0.48	0.5	F = 0.08	0.78
Left main pulmonary artery inner diameter / right main pulmonary artery inner diameter	0.94	0.92	F = 0.06	0.81
CT value of ascending aorta (HU)	414	289	F = 1.60	0.24
CT value of descending aorta (HU)	347.83	394.75	F = 5.11	0.05
CT value of ascending aorta / CT value of descending aorta	1.17	0.75	F = 0.62	0.46

3.4 Surgical findings

A total of 10 patients underwent surgical treatment and DSCT reexamination within 3 months after the operation, including 1 case of type A and 9 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 that of 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 was restored postoperatively.

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

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 line, 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. Making the ligation of the arterial catheter, cutting the pulmonary artery, and using 4 – 0 prolene double-ended needle with a gasket to close the arterial catheter, and then pulmonary artery was formed. After rewarming, fully exhausting, and opening the ascending aorta, the heart automatically returned to beating, sinus rhythm, and smooth shutdown. 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 aortoplasty. The artificial blood vessel was constricted, and the other end of the anastomosed artificial blood vessel was pulled into the pericardial cavity through the left pulmonary hilum over the top of 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 4 – 0 Prolene line for end-to-side anastomosis. After rewarming, adequate exhausting, the opening of ascending aorta, the heart resumes automatically, sinus rhythm.

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

4 Discussion

IAA is a rare congenital cardiovascular malformation, accounting for 1% of the critical congenital heart diseases in children [11–13]. The incidence rate of IAA is about 0.19/1000 live births, accounting for 5% of the cases of aortic arch obstruction [14, 15]. The formation of IAA is related to chromosomal recombination and single gene abnormality [16].

The formation of IAA is related to chromosome recombination and single gene abnormality [16], 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 2 cases (8.00%) were not accompanied by other heart diseases, and the IAA patients accompanied with ventricular septal defect and patent ductus arteriosus (9 cases, 36%) were the most, which was basically close to the above study [16]. IAA was first described in 1778, and has 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 [12–13]. In this study, 10 patients completed surgical treatment, and all patients survived.

4.1 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 [17]. The degree of collateral circulation in the chest, abdomen, and lower limbs supplied by the descending aorta is the key to the patients' 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 [18]. In this study, all adult patients had abundant collateral circulation. However, there were three patients in the study that 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 about 1-year-old child. The clinical manifestations of adult IAA are diverse, including asymptomatic, headache, hypertension, and differences in blood pressure between upper and lower limbs [19]. 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.

4.2 Classification characteristics of IAA

IAA is characterized by anatomical discontinuity between 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. [10]. 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, which is the interruption of the aortic arch between the brachiocephalic trunk and the opening of the left common carotid artery, caused by the abnormal degeneration of the left third and fourth arch arteries [20–21]. Compared to the study by Fournier et al., there are more patients 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 deviation. Therefore, more studies are required to improve the epidemic-related data of this disease. Arentje et al. [8] further divided IAA into 6 subtypes according to different sites of interruption and opening position of the right subclavian artery. However, the descending aorta originated from pulmonary artery. In their study, 35% of IAA were type A and 6% were type A1, while in our study, 15 (60%) cases of type A were severed at the distal end of the left subclavian artery orifice, and 5 (20%) cases of type B were located between the left common carotid artery and the left subclavian artery orifice, while in the study by Arentje et al., 53% were type B and 2 cases were type B1 (8%). The transection 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% cases were type B1. In our study, it is rare that the type C is not observed clearly. It is located between the innominate artery and the left common carotid artery. Among them, 3 (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. [9]. Because the source of blood supply in the descending aorta is related to cyanosis, Wang et al. classified IAA into 5 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% 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, 1 (4.00%) case did not belong to the above classification. The interruption of the 1 case occurred between the left common carotid artery and the left subclavian artery opening, and the descending aorta is supplied by the thoracic collateral branch circulation (Fig. 3). Therefore, the author suggests adding a type G to the "two types and five types"(Fig. 3A) classified on the basis of cyanosis and non-cyanosis modified by Wang et al. [9], 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 described previously.

4.3 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 basically replace the invasive digital subtraction angiography (DSA) examination [22]. 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 [17] reported that IAA is increased, as observed by CTA[1]. 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 patients with 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 author 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 type A The patient's pulmonary artery has a wider inner diameter. There is no difference between of 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 can also 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, the celiac artery, the left hepatic artery, the internal iliac, and the external arteries. There are also 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 [23]. 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 aorta [24].

4.4 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 [25]. Whether IAA adults need surgical treatment is controversial, which needs to be combined with the patient's condition [26]. Because the adult patient survivors have abundant collateral circulation compensation, some patients can survive without surgery and can be treated conservatively.

4.5 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 [27]. 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 [28].

5 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.

6 Conclusion

In conclusion, IAA is a rare congenital malformation. In summary, 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 ascending aorta, descending aorta, 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 in diagnosing IAA.

Abbreviations

AAT: Aortic arch thrombosis; CPB: Cardiopulmonary bypass; COA: Coarctation of the aorta; CT: Computed tomography; CTA: Computed tomography angiography; CPR: cerved projection reformation; DBP: Diastolic blood pressure; DSA: Digital subtraction angiography; DSCT: Dual-source computed tomography; HU: Hounsfield unit; IAA: Interrupted aortic arch; MR: Magnetic resonance; MPR: Multiplanar reorganization; PDA: Patent ductus arteriosus; PACS: Picture Archiving and Communication Systems; SBP: Systolic blood pressure; VSD: Ventricular septal defect; VR: Volume rendering.

Declarations

Acknowledgements

Authors' contributions

Yusen Feng and Lijuan Wang made substantial contributions to the conception and design of the work; Hui Liu and Guifang Sun made substantial contributions to the acquisition and analysis; Bin Liu made substantial contributions to interpretation of data; Liqiong Wang and Xuguang Zhang made substantial contributions to the creation of new software used in the work; Yusen Feng and Pengcheng Ma made substantial contributions to draft the work and substantively revise it.

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Availability of data and materials

The datasets used and/or analysed during the current study are available from the corresponding author on reasonable request.

Ethics approval and consent to participate

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.

Consent for publication

Not applicable.

Competing interests

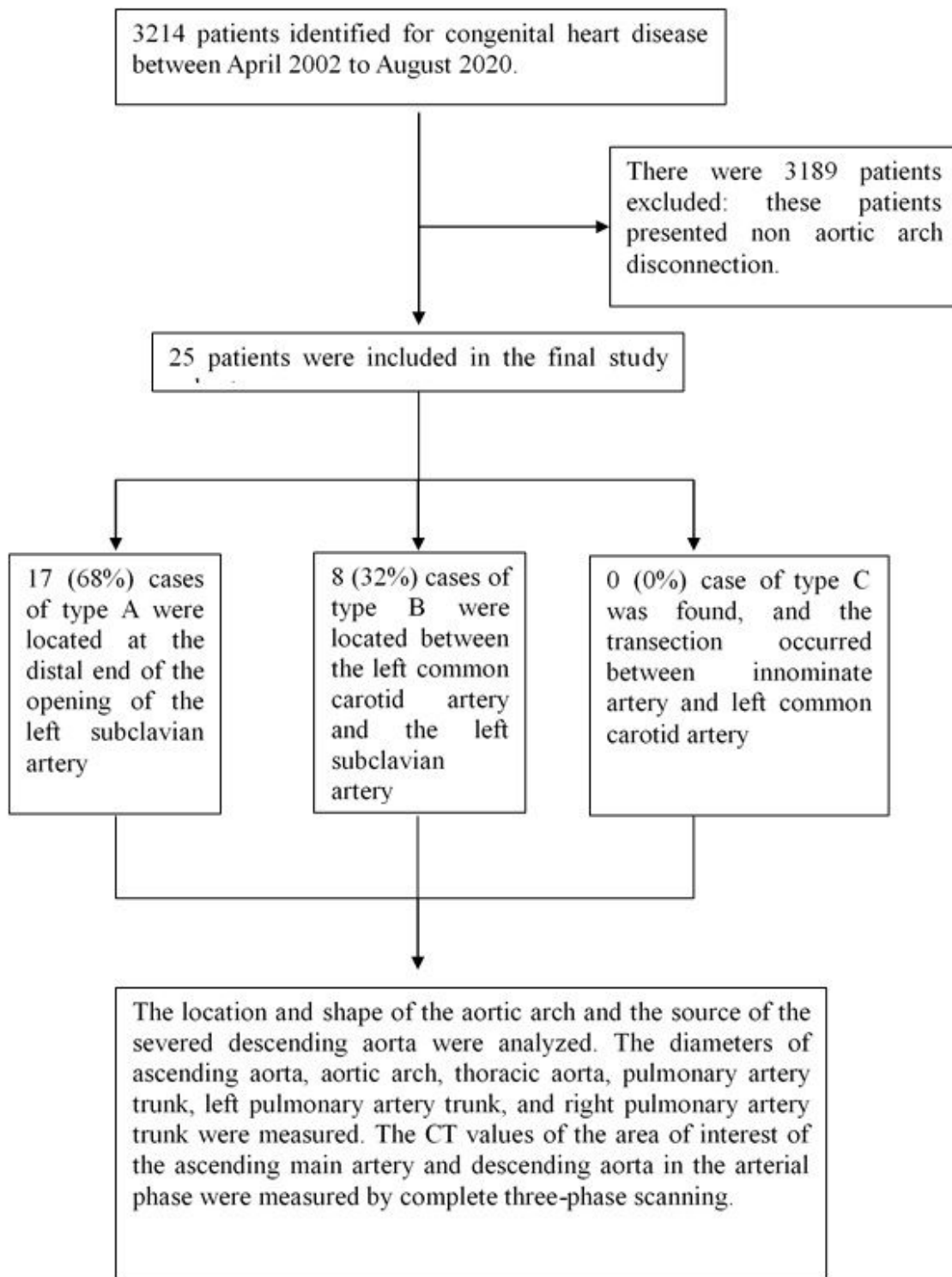The authors declare that they have no competing interests.

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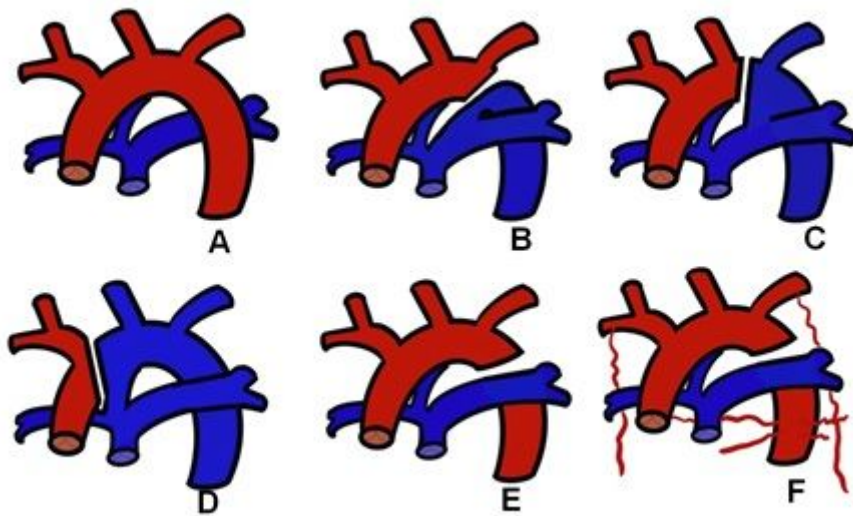
Figures

3214 patients identified for congenital heart disease between April 2002 to August 2020.

There were 3189 patients excluded: these patients presented non aortic arch disconnection.

25 patients were included in the final study

17 (68%) cases of type A were located at the distal end of the opening of the left subclavian artery

8 (32%) cases of type B were located between the left common carotid artery and the left subclavian artery

0 (0%) case of type C was found, and the transection occurred between innominate artery and left common carotid artery

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 were 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.
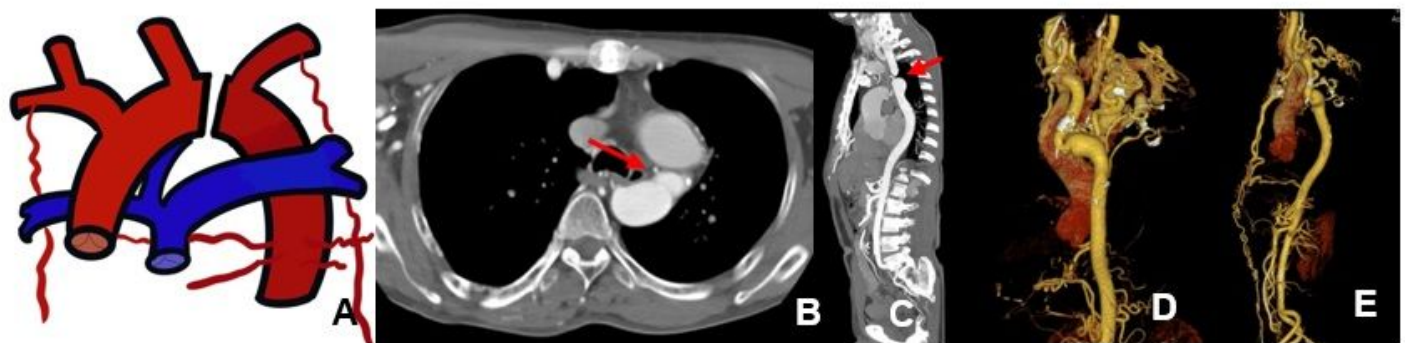
Figure 1

flow diagram.

Figure 2

IAA classification diagram. (A) is a schematic diagram of a normal aortic arch; (B) is a 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) is a 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 is often combined with the right subclavian artery and often vagus, and the descending aorta is supplied by the pulmonary artery; (D) is a 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) is a 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) is a 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) is 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) shows the transverse section image of CT vessel enhancement: the aortic arch and descending aorta are severed (pointed by the long red arrow); (C) shows the sagittal image of the CT vascular enhancement: the aortic arch and descending aorta are severed (pointed by the short red arrow); (D and E) are three-dimensional reconstruction images of blood vessels: There is no direct blood flow communication between the left common carotid artery and the 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 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 right axillary artery communicated with the right expanded intercostal artery.