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**Name of Journal:** *World Journal of Clinical Cases*

**Manuscript NO:** 69792

**Manuscript Type:** CASE REPORT

**Difference and similarity and connection between type A interrupted aortic arch and aortic coarctation in adults: Two case reports**

Ren SX *et al.* The IAA and CoA in adults

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**Abstract**

**BACKGROUND**

Aortic coarctation (CoA) is usually confused with interrupted aortic arch (IAA), especially adult type A interrupted aortic arch due to their similar anatomical location. Although the main difference between them is whether arterial lumen exhibits continuity or not, the clinical manifestations are similar and connection exists between them. Adult type A IAA is considered as an extreme form of CoA, which is complete discontinuity of aortic function and lumen caused by degenerative arterial coarctation. This paper reports two cases (interrupted aortic arch and severe aortic coarctation) to analyze the difference and similarity and connection between them.

**CASE SUMMARY**

The two cases of patients presented with hypertension for many years. Computed tomography angiography (CTA) showed that aortic arch and descending aorta were discontinuous or significantly narrowed with extensive collateral flow. The IAA patient refused surgical treatment and blood pressure could be controlled by drugs. While the CoA patient underwent stent implantation because of uncontrollable hypertension, the

blood flow recovered smoothly and the blood pressures at both ends of the stenosis returned to normal after surgery.

## CONCLUSION

Adult type A IAA and CoA have difference and similarity, and type A IAA is associated with CoA to a certain extent. Choose the treatment method based on the patient's clinical symptoms rather than the severity of the lesion.

**Key Words:** Interrupted aortic arch; Aortic coarctation; Computed tomography Angiogram; Case report

Ren SX, Zhang Q, Li PP, Wang XD. Difference and similarity and connection between type A interrupted aortic arch and aortic coarctation in adults: Two case reports. *World J Clin Cases* 2022; In press

**Core Tip:** According to the characteristics of IAA and CoA, this paper highlights the difference and similarity and connection between them. Choose the treatment method based on the patient's clinical symptoms rather than the severity of the lesion.

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## **INTRODUCTION**

Interrupted aortic arch (IAA) is a congenital malformation disease with loss of continuity between aortic arch and descending aorta, accounting for about 1% of congenital heart disease<sup>[1]</sup>. According to the location of the dissection, it is divided into three types<sup>[2]</sup>: the dissection is located at the distal to the left subclavian artery origin (type A); between the left common carotid artery and the left subclavian artery (type B); between the brachiocephalic trunk and the left common carotid artery (type C). Type A accounts for approximately 79% of adult IAA<sup>[3]</sup>, without the complications associated with type B such as ventricular septal defects and patent ductus arteriosus<sup>[1,4-6]</sup>. Coarctation of the aorta (CoA) is defined as a discrete stenosis of the aorta usually located in the area of the ligamentum arteriosum, with a higher incidence of 6%-8% than IAA<sup>[3]</sup>. To investigate the characteristics if there were difference, similarity and connection in adult type A IAA and CoA, cases of both types were reported in this study. The patients had provided informed consent for publication of the cases.

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## **CASE PRESENTATION**

### ***Chief complaints***

A 54-year-old woman presented to the hospital with chief complaints of discomfort in right waist and abdominal pain for a week. And a 61-year-old female patient, with tiredness and dizziness, was diagnosed with CoA during physical examination a year ago.

### ***History of present illness***

Her medical history was notable for hypertension and the right renal cyst in case 1. And the described patient in case 2 was diagnosed with CoA during physical examination a year ago with tiredness and dizziness.

### ***History of past illness***

The first woman in case 1 indicated hypertension for 8 years. The second patient in case 2 with a history of hypertension for 30 years had the highest blood pressure - 190/100 mmHg, and a clear medical history of thyroidectomy and coronary artery stenosis.

### *Physical examination*

In case 1, continuous murmurs were heard on the chest and back during auscultation. The blood pressure was monitored, after taking the medication the left upper and left lower limbs blood pressures were 145/77 mmHg and 120/64 mmHg respectively. The dorsalis pedis were thin and weak. In case 2, the upper limbs pressures were higher than lower limbs pressures.

### *Laboratory examinations*

Laboratory examination indicated the following in case 1: uric acid, 412  $\mu\text{mol/L}$ ; triacylglycerol 1.95mmol/L; glucose, 5.5 mmol/L.

Laboratory examination indicated the following in case 2: urea, 3.25 mmol/L; creatinine, 50.8  $\mu\text{mol/L}$ ; cholesterol, 3.94 mmol/L; alanine aminotransferase, 19 U/L; aspartate aminotransferase, 23 U/L; glucose, 4.81 mmol/L.

In the described patients, Urinalysis, coagulation tests, complete blood count, and infection marker tests did not reveal any obvious abnormalities.

### *Imaging examinations*

**Case 1:** Electrocardiogram (ECG) showed complete right bundle branch block and T wave change. Color doppler echocardiography displayed normal right atrium and right ventricle, enlarged left atrium and left ventricle. The aortic arch and the distal to the left subclavian artery were discontinuous, and numerous collateral branches existed around the descending aorta. The flow of abdominal aorta showed small slow wave.

Abdominal computed tomography (CT) examination revealed multiple tortuous vascular shadows in the paravertebral column and bilateral chest wall areas. To further clarify the diagnosis, thoracic and abdominal computed tomography angiogram (CTA)

showed that aortic arch and descending aorta were discontinuous, and the disconnection was located at the distal to the left subclavian artery (type A IAA) (Figure 1A). The ascending aorta was dilated with normal curvature and aneurysm formation in descending aorta (Figure 1A and B). Bilateral internal mammary arteries, intercostal arteries, thoracoabdominal wall arteries and multiple arterial branches tortuously expanded to supply blood to distal vessels and organs. For example, the bilateral internal mammary arteries were connected to the external iliac arteries (Figure 1C), and the blood of the abdominal trunk came from the intercostal artery and the inferior phrenic artery.

**Case 2:** ECG showed first degree atrioventricular block and abnormal Q wave in V1-V3. Color doppler echocardiography indicated that no obvious enlargement in the atria and ventricles of the heart.

CTA examination showed that the lumen of the aortic isthmus was significantly narrowed (Figure 2A), the normal curvature of the ascending aorta was maintained, and the proximal descending aorta was slightly enlarged (Figure 2A). The bilateral internal mammary arteries were connected with the distal external iliac arteries (Figure 2B).

### ***Medication History***

The maximum systolic pressure was acceptably controlled in the past by Valsartan and Hydrochlorothiazide Tablets and Levamlodipine Besylate Tablets in case 1. Oral administration of Nifedipine Controlled-release Tablets (Bai Xintong), Candesartan Cilixetil and Indapamide Sustained Release Tablets did not control blood pressure well in case 2.

### **FINAL DIAGNOSIS**

Based on the images of Figure 1 and 2, we diagnosed the patient as type A IAA and CoA with extensive collateral flow, respectively.

## **TREATMENT**

Without other positive symptoms, the first patient only required treatment of the relevant symptoms. The blood pressure controlled by antihypertensive drugs was acceptable during the follow-up period.

Due to uncontrollable hypertension, the second patient was admitted to the hospital and received aortic angiography and stent implantation under general anesthesia. The angiography indicated severe narrowing of the descending aorta at the distal to the left subclavian artery at 15 mm, with a length of 10 mm. Before stent placement, the blood pressures at both ends of the constriction were 185/73 mmHg and 110/65 mmHg, respectively. After stent placement, the blood flow recovered smoothly, and the blood pressures at both ends of the stent were 127/62 mmHg and 122/62 mmHg respectively.

## **OUTCOME AND FOLLOW-UP**

Both them have remained stable.

## **DISCUSSION**

The difference between type A IAA and CoA in adults is the absence or presence of continuity of the arterial lumen. As shown in case 1, there was no anatomical continuity between the distal to the left subclavian artery and descending aorta (type A IAA). In contrast, no obvious anatomical interruption was detected in CoA case with the narrowing between the above-mentioned arteries. According to previous reports, although the definitions of type A IAA and CoA were different, the clinical manifestations were similar in increased blood pressure which upper limbs pressures were higher than lower limbs pressures<sup>[7-12]</sup>. The manifestations of case 1 and 2 in this study were consistent with these reports. The pathophysiological changes are: first, imbalance distribution of blood flow and blood pressure in the upper and lower limbs is caused by increased mechanical resistance resulted from interruption or stenosis of

the arterial lumen; second, hypoperfusion in lower bodies results in the activation of renin-angiotensin-aldosterone system<sup>[12]</sup>. Typical symptom was not observed in the lower extremities of two cases due to extensive collateral circulation was established to supply blood flow to the distal organs in case 1 and 2. As aggravation of the narrowing, upper limbs blood pressures can be significantly increased, which may cause a series of symptoms such as upper limbs pain, headache, dizziness, angina, lower limbs chills and claudication.

The mechanism of IAA in adult is different from that in infancy<sup>[1]</sup>. The most common type in infancy is the type B IAA which is closely related to congenital developmental abnormalities and may be concurrent with other anomalies including patent ductus arteriosus, aneurysm, ventricular septal defect, *etc*<sup>[1,9]</sup>. While type A IAA in adult is similar to CoA in diseased anatomical position which is located between the left subclavian artery and the ductus arteriosus<sup>[13]</sup>. Type A IAA in adult may be the result of regression or atrophy of the diseased arterial segment as this segment can be narrowed, atretic, fibrotic, or completely absent<sup>[2]</sup>. Some researchers<sup>[9,13]</sup> believed that in CoA, the ascending aorta maintained normal curvature, the narrowing was located several centimeters away from the left subclavian artery, and distal to the coarctation was enlarged or formed aneurysm because increased velocity of blood flow at the region damaged the vessel wall<sup>[6,14]</sup>. These findings were seen in Case 1 which strongly suggested that adult type A IAA is the final stage of aortic coarctation degeneration. The IAA cases caused by severe CoA have also been reported in the literature<sup>[15,16]</sup>, the patients initially showed aorta narrowing. Over time, the complete obstruction of the narrowed lumens led to aortic interruption, which was eventually treated by surgical intervention.

The blood pressure of the patient with type A IAA in Case 1 could be controlled by drugs, so surgical intervention was not performed. Generally, the main treatment for IAA is to reconstruct the blood flow at both ends of the interrupted arteries, which can be achieved by surgical means (such as end-to-end anastomosis, graft interposition, or extra-anastomotic bypass) and percutaneous approach<sup>[1]</sup>. At present, there is still a lack



of long-term follow-up data on adult patients with type A IAA repaired, and postoperative complications in these patients require continuous attention. The purpose of surgery in CoA is the same as in type A IAA, which is to reestablish the lesion to enable appropriate blood flow. Surgical methods for the treatment of CoA are mainly balloon angioplasty and stent placement<sup>[17]</sup>. The patient with severe CoA in case 2 was treated with stent placement because of uncontrollable hypertension. Blood pressures at both ends of the stenosis returned to normal after surgery, and long-term follow-up observation will be essential for a series of complications after CoA is repaired, such as restenosis, hypertension, coronary artery disease, aortic rupture, and aneurysm formation<sup>[3,11,17]</sup>.

### **CONCLUSION**

Adult type A IAA and CoA have difference and similarity. The difference lies in the absence or presence of continuity of the arterial lumen, while the similarity is that they are accompanied by secondary hypertension with extensive collateral circulation. In conclusion, type A IAA is associated with CoA to a certain extent, which can be considered that the arterial lumen is completely interrupted between aortic arch and descending aorta with the degeneration or atrophy of arterial narrowing.

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SIMILARITY INDEX

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