Is surgery necessary for adults with Isolated Interrupted Aortic Arch?: Case series with literature review

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Abstract

Background and Aim: Interrupted aortic arch (IAA) is defined as a complete interruption of aortic lumen between the ascending and descending aorta. It is an uncommon and complicated congenital heart disease with high mortality in infants. It is rare for patients with isolated IAA to survive to adulthood without operation unless the extensive collateral vessels joining the descending aorta. Here we present three unique cases with isolated IAA together with a review of the literature. Methods: case presentation: We retrospectively searched the hospital patient databases for patients (>14 years) with IAA diagnosed at the Wuhan Union Hospital over the past 10 years and excluded the patients with other cardiac malformations. Three patients were identified. Two were referred to us for hypertension management and were diagnosed with IAA at our hospital. They both declined surgical treatments and underwent conservative therapy including management of their hypertension. One patient was referred to our hospital for further treatment options after the patient was diagnosed with IAA at another hospital. This patient received an extra-anatomic bypass surgery from ascending aorta to descending aorta. His high blood pressure did not resolve and was subsequently managed by anti-hypertensives medications after the surgery. Discussion and conclusions: Adult patients with isolated IAA usually have extensive collateral vessels joining the descending aorta. Surgical intervention may not be necessary for these patients if the patients have no symptoms except hypertension. Anti-hypertensives medical management with long term follow-up appears to be a reasonable treatment option for these patients.

Introduction:

Interrupted aortic arch (IAA) is a rare congenital aortic anomaly due to a complete interruption of aortic lumen between the ascending and descending aorta [1]. It has a high mortality in infants accounting for 1% of congenital cardiovascular diseases. Approximately 75% of patients with IAA died by 10 days after birth and 90% died within the first year after birth [2]. Based on the presence or absence of other associated deformities, IAA can be categorized as complex and isolated types. The former is more common and usually associated with ventricular septal defect and patent ductus arteriosus [3,4].The latter is rare, and patients with isolated IAA rarely survive to adulthood without surgical intervention unless the extensive joining of the collateral vessels to the descending aorta [5].Patients receiving operations in infancy were in diminished health status with significant diminished CPET performance and preserved resting LV function.The operative re-intervention rates was 29% and 18% required LVOT interventions over 15 years of follow-up [6].In this article, we will present three cases of adults with isolated interrupted aortic arch, and discuss about the theupatic strategy that whether surgical intervention is the best option for adult patients.

Materials and Methods:

Case presentation: Hospital database was searched for adult patients who had a discharge diagnosis of IAA at Wuhan Union Hospital over the past 10 years based on the international classification of diseases codes. The clinical notes of each potential case were reviewed, and patients younger than 14 years old or with other

cardiac malformations were excluded. We also conducted a comprehensive literature search regarding IAA. Wuhan Union Hospital institutional review board approved this study.

3 patients were identified who were diagnosed with IAA at our institution. They were admitted to our department with the chief complaint of hypertension and later diagnosed with IAA using computated tomography angiography(CTA) or transcatheter angiography. Besides hypertension, they have no other symptoms. Two patients declined surgical treatment and received conservative therapy including anti-hypertensive treatment. The third patient received an extra-anatomic bypass surgery from ascending aorta to descending aorta. His post-surgical blood pressure remained high and was treated with irbesartan and hydrochlorothiazide.

Case 1:A 43-year-old man was admitted to our department with the complaint of high blood pressure in 2020. Physical examination showed regular heart beat with blood pressure of 179/102 mmHg in the right arm, 170/100 mmHg in the left arm, 135/80 mmHg in the right limb and 125/70 mmHg in the left limb, and there was a systolic murmur around the sternum. Electrocardiogram showed sinus rhythm. Echocardiography showed possible coarctation of aorta without other cardiac malformations. Then he was referred to the radiology department to further evaluate a possible aortic abnormality. CTA showed an interruption of the aortic arch distal to the origin of the left subclavian artery(Type A, Fig1 A and B) and profuse collateral circulation. The blood biochemistry laboratory results showed normal renal and liver function. There were no signs or symptoms of ischemia of splanchnic organs. Since the patient had no symptoms other than hypertension, we recommended anti-hypertensive management. After taking valsartan amlodipine and felodipine, his daily blood pressure in the left arm was stabilized at about 140/90 mmHg.

Case 2:Another 43-year-old man was admitted to our department for further treatment after he was diagnosed with IAA in another hospital in 2017. His physical examination showed regular heart beat with blood pressure of 132/96 mmHg in the right arm, 130/92 mmHg in the left arm, 110/80 mmHg in the right limb and 108/70 mmHg in the left limb, and there was a systolic murmur around the sternum. Before his referral, he had had high blood pressure for one year with the highest systolic pressure in the left arm up to 180 mmHg. Before he came to our department, he had already been diagnosed with IAA by another hospital and had been taking anti-hypertensive drugs for several weeks. His electrocardiogram showed in sinus rhythm with incomplete right bundle branch block. His echocardiography indicated possible coarctation of aorta with widened STJ (sinotubular junction). CTA revealed a coarctation or interruption of descending part of aortic arch with ascending aortic aneurysm and abundant collateral circulation. To determine whether it was a coarctation or an interruption, we performed an aortic angiography via right femoral artery and right radial artery, confirming that it was an interruption of the aortic arch distal to the origin of the left subclavian artery (Type A) with profuse collateral circulation(Fig1 C and D). His blood biochemistry laboratory results showed normal renal and liver function, without signs of ischemia of splanchnic organs. Similar to case 1, we recommended anti-hypertensives with perindopril, and the patient's daily blood pressure in the left arm has stabilized at about 130/80 mmHg.

Case 3:A 28-year-old man was admitted to our department with the complaint of high blood pressure in 2017. Physical examination showed irregular heart beat with blood pressure of 174/110 mmHg in the right arm, 180/108 mmHg in the left arm, 130/82 mmHg in the right limb and 135/84 mmHg in the left limb, and there was a systolic murmur around the sternum. Electrocardiogram showed in sinus arrhythmia. Before he came to our hospital he had had echocardiography and CTA. His echocardiography showed coarctation of the descending aorta without other cardiac malformations, and CTA showed coarctation of the descending aorta with abundant collateral circulation. Initially, we attempted to place a stent via aortography. However, the aortography via right femoral artery and right radial artery showed an interruption of the aortic arch distal to the origin of the left subclavian artery (Type A) with abundant collateral circulation(Fig1 E and F). The blood biochemistry results showed normal renal and liver function without signs of ischemia of splanchnic organs. Since the patient had no symptoms other than high blood pressure, as case 1 and 2, we recommended anti-hypertensives. However,given this patient's young age and concerning regarding possible future IAA-related complications such as heart failure, renal and liver damage, etc., the patient and his family requested a surgery to correct the aortic deformities. We performed a surgical reconstruction to

restore the continuity of aortic arch using extra-anatomic bypass from ascending aorta to descending aorta with 16mm artificial blood vessel. The surgery went well, but the patient's systolic pressure in the right arm remained at approximately 170 mmHg. He received anti-hypertensives and accomplished satisfactory blood pressure (about 140/90 mmHg in the left arm), which remained adequate, 2 years after the surgery. And the patient was comatose for 2 weeks after the surgery. Since then he had been with right limb weakness and suffered from epilepsy. Significant diminished CPET performance and decreased communication ability accompanied. He could only walk slowly for about 50 meters and have a simple chat with others.

Discussion and Conclusion:

Interrupted aortic arch (IAA) is caused by a complete interruption of aortic lumen between the ascending and descending aorta. It is a rare and complicated congenital cardiovascular condition with high mortality in infants. Approximately 75% of patients with IAA died by 10 days after birth and 90% died within the first year after birth[2]. Steidele, *et al.* reported the first case of IAA in 1778 [7] and the the first surgical correction of IAA was performed by Samson in 1955 [8].

Based on the position of the interruption, IAA can be clinically categorized into 3 types(1). Type A(40%): the aortic arch interruption is at the distal end of the left subclavian artery. Type B(55-60%): the interruption is between the left subclavian artery and the left common carotid artery and strongly associated with DiGeorge syndrome and chromosome 22q11.2 deletion [9,10]. Type C(4%): The interruption is between the innominate artery and left common carotid artery. Patients with Type A IAA are more likely to have adequate collateral flow and thus are commonly asymptomatic. Type B and C IAA have disparate upper extremity pressures making diagnosis in childhood and adolescence more common. Based on associated additional deformities, they can be further classified as complex and isolated types. Complex types are common and usually occurs with other cardiac malformations like patent ductus arteriosus (PDA), aortopulmonary window, ventricular septal defect (VSD), etc. [3,4]. These malformations can guarantee the blood supply of descending aorta. Isolated type is very rare [11].

Because IAA always combines with PDA and VSD, some newborns seldom have overt heart symptoms in early weeks after birth. However, at a later phase of the infancy, the decreasing of pulmonary vascular resistance and increased left to right shunt can cause congestive heart failure. In many cases, the ductus arteriosus begins to close shortly after birth. If the ductus arteriosus closes too rapidly, the collateral circulation between the ascending and descending aortas is not well developed. Consequently poor perfusion of the lower part of the body can lead to ischemia of splanchnic organs, severe acidosis, and anuria[4,7]. Therefore, it is rare for patients with isolated IAA to survive to adulthood without surgical intervention unless the extensive collateral vessels joining the descending aorta are established [11].

Most of patients with isolated IAA in adulthood come to hospital with the complaint of hypertension [12]. Other common symptoms include claudication and congestive heart failure[13-15]. It is difficult to identify such patients unless discrepancy between the blood pressure of the upper and lower extremities is found or echocardiography is performed. The pulsation of lower or up extremity artery may be helpful but it depends on the position of the interruption.

Echocardiography is a useful initial screening method, particularly important for neonates, to identify and measure necessary anatomical data of IAA, and identify other cardiac malformations[2,16]. CTA provides a high resolution 3-dimensional image of IAA and collateral vessels [12,17,18]. The combination of echocardiography and CTA is usually recommended for a definitive diagnosis [9]. Although aortography can demonstrate complete occlusion of the aorta and the position of the interruption, it is usually not the first choice due to its invasive procedure. Nevertheless, an aortography is necessary when CTA can not distinguish between aortic arch interruption and coarctation.

The main treatment for IAA in neonates and infants is surgical intervention [15,19]. There is a growing preference for a radical surgery which includes a reconstruction of aortic arch continuity and repair of cardiac malformations under deep hypothermic circulatory arrest in a single-staged procedure. Several surgical means, such as end-to-end or end-to-side anastomosis can be used[15]. For neonates and infants,

blood can flow through the artery catheter into the descending aorta. Before operation, prostaglandin E can be used intravenously, with endotracheal intubation and mechanical ventilation support. High tidal volume and high concentration of oxygen supply should be avoided since they may cause respiratory alkalosis.

The main treatment for IAA in adults is the same with in neonates or infants. About 90% of patients prefer a surgical correction. Either sternotomy or thoracotomy seems reasonable depending on the size and location of collaterals. However, end-to-end or end-to-side anastomosis is less common in adults than the extra-anatomic bypass[20,21]. When collateral circulation is abundant, surgery can be difficult due to high bleeding risk [21]. Krishna *et al.* reported one case that one patient received subclavian to descending aorta bypass required surgical re-exploration 3 times for bleeding from ruptured collaterals [22]. Freedman H K *et al* first proposed the theory that some type A IAA may be the result of regression or atrophy of a previously existing segment of aortic arch between the ductus arteriosus and left subclavian artery comparable with coarctation of the aorta as this segment can be narrowed, atretic, and replaced by a fibrous band, or completely absent[23]. Kusa *et al.* cautioned that the percutaneous stent placement was possible in those patients. He also cautioned that immediate access to cardiac surgery must be available in case of an emergency [7,14].

In most of reported cases, patients' symptoms intend to improve after surgery. Hypertension gradually resolves after surgery, and anti-hypertensive management can be discontinued. Other symptoms such as claudication and paresthesia are also ameliorated after surgery[9,24]. However C. Sai Krishna *et al* reported that among seven patients received operation, five patients still had residual mild hypertension that was well-controlled with anti-hypertensive management [21]. Similarly in our case 3, the patient's blood pressure remained high after the surgery. Considering that IAA is similar to coarctation of the aorta (CoA) , we reviewed the relevant data of CoA and found that 68% of patients still have hypertension after surgery. This lack of blood pressure improvement may be related to abnormal geometry of the aortic arch or hypoplasia of the arch changes in arterial function, hyperactivation of the renin angiotensin system, and attenuation of baroreflexes[24]. So for patients with abundant collateral and no symptoms of ischemia of splanchnic organs and lower limbs, anti-hypertensive management without surgical intervention is reasonable. Nonetheless, close follow-up and further study is necessary.

Although it is extremely uncommon, IAA must be considered in the diagnosis of an adult patient with severe hypertension. For adult patients with isolated IAA and no symptoms besides hypertension, it likely that their collateral circulation is sufficient. For such patients, surgical intervention may not be the best option,while anti-hypertensives management appears to be adequate and safe.

Abbreviations:

IAA: Interrupted Aortic Arch

LV: Left ventricular

LVOT: Left ventricular outflow tract

CPET: Cardiopulmonary exercise testing

CTA: Computated tomography angiography

STJ: Sinotubular junction

PDA: Patent ductus arteriosus

VSD: Ventricular septal defect

CoA: Coarctation of the aorta

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Figure Legends:

Figure 1:

A and B:Case1, CTA revealed a type A interrupted aortic arch with complete discontinuity of the aortic lumen distal to the origin of the left subclavian artery

C and D:Case 2, angiography showed absence of anatomical continuity between the ascending aorta distal to the left subclavian artery and the descending aorta.

E and F:Case 3, angiography showed absence of anatomical continuity between the ascending aorta distal to the left subclavian artery and the descending aorta.