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Medical Treatment of an Adult with Uncorrected Isolated Interrupted Aorta Resulted in no Complications after 4 Years of Follow-Up

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

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Conflict of interest: None declared

Patient: Female, 56
Final Diagnosis: Isolated adult interrupted aortic arch
Symptoms: Headache • hypertension • left ventricular hypertrophy
Medication: —
Clinical Procedure: —
Specialty: Surgery

Objective: Congenital defects/diseases

Background: Interrupted aorta is a rare congenital malformation defined as the lack of continuity between the ascending and descending parts of the aorta.

Case Report: This malformation was first described by Steidle in 1778. To date a few isolated adult interrupted aortic arch patients have been reported and most of them were treated surgically. However, there is not data about outcome of patients who decline surgery or who are not good candidates for surgery because of excessive risks, and there is not a data about how to follow these patients.

Conclusions: Herein we present a case of adult type A isolated interrupted aorta followed-up for 4 years by medical therapy without complications.

MeSH Keywords: Aorta, Thoracic • Imaging, Three-Dimensional • Interdisciplinary Communication

Full-text PDF: <http://www.amjcaserep.com/abstract/index/idArt/890716>



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Background

Interrupted aorta, which is defined as lack of continuity between the ascending and descending parts of the aorta, is a rarely encountered clinical problem in adulthood [1]. This malformation was first described by Steidele in 1778 [2].

Interrupted aorta is encountered in 3 of 1 million live births, and constitutes 1% of all congenital cardiac malformations [3]. Types of this malformation were first described by Celoria and Patton in 1959 according to interruption site [4].

Type A constitutes 43% of all patients, and the lesion site is distal to the left subclavian artery. Type B is the most common type and constitutes 53% of all patients, and the lesion site is between left common carotid artery and left subclavian artery. The rarest type is C, in which the interruption is between the innominate and left carotid artery. Multiple VSDs, truncus arteriosus, left ventricular outflow obstruction, valvular aortic stenosis, transposition of great arteries, double-outlet right ventricle, and aorto-pulmonary window can accompany the interrupted aorta.

The only described treatment option in the literature is surgery [5,6]. However, some patients might reject surgery and some are not eligible for surgery.



Figure 1. Sagittal oblique reformatted CT angiography image showing the interrupted aortic arch distal to the left subclavian artery.

There is an absence of data on how to manage these patients. Here, we describe an adult interrupted aorta case followed successfully by medical therapy for 4 years.

Case Report

In September 2009, a 56-year-old woman was admitted to the hypertension clinic in our institution. Her only complaint was a headache at the back of her neck, associated with hypertension. On admission physical examination, blood pressure was 170/110 and heart rate was 92 bpm. Her blood pressure was the same at both arms. Development imbalance was not observed in her body structure, and her lower extremity pulses were not weak, but lower extremity blood pressure measurements revealed 150/90mmhg, which is below the upper limit of values. On the cardiac auscultation, a harsh 3/6 systolic murmur, which was loudest at the left scapular region, was heard. The other system examination results were normal.

The ECG showed a typical left ventricular hypertrophy pattern. On transthoracic echocardiogram (TTE) we observed concentric left ventricular hypertrophy and mild aortic insufficiency, depending upon the degenerative changes of aortic cusps. Due to poor image quality at the suprasternal window, we could not clearly evaluate the gradient in the descending aorta.

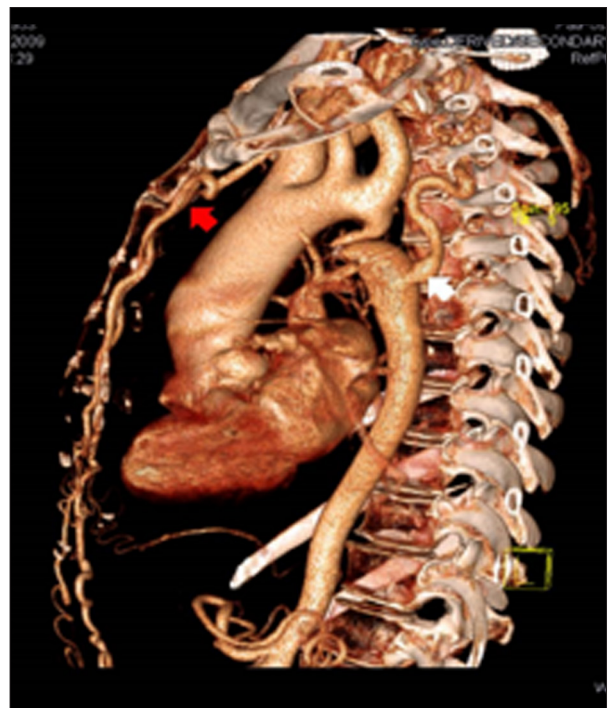


Figure 2. Volume-rendering CT image showing extensive collaterals. Dilated internal mammary arteries (red arrow) and enlarged intercostal arteries (white arrow) are seen.

Because of unexplained murmur and inadequate suprasternal image quality, thorax computed tomography (CT) angiography was planned to exclude an aortic malformation.

Multidetector CT angiography revealed interrupted aortic arch distal to the left subclavian artery. Dilated internal mammary arteries and enlarged intercostal arteries were the major collaterals supplying blood to the distal part of the aorta (Figures 1 and 2).

According to the few reports regarding isolated interrupted aortic arch, we recommended surgery to the patient [5,6], but the patient declined the operation because of surgical risks. According to the current literature, there is insufficient data and no clear consensus about following up these kinds of patients.

Therefore, we arranged a surveillance team consisting of a cardiologist, a radiologist, a neurologist, and an ophthalmologist. We prescribed Irbesartan/Hydrochlorothiazide (300/12.5) nebivolol 5 mg to the patient. At the first-year follow-up, we had to add lercanidipine 10 mg because of insufficient control of the hypertension. The surveillance team examined the patient at regular intervals. We could not do follow-up CT angiography because she declined it. After 4 years of follow-up, her blood pressure is under control and she is free of complaints and complications.

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Discussion

Interrupted aorta is a rare congenital malformation defined as the lack of continuity between the ascending and descending parts of the aorta.

Interrupted aortic arch is a fatal condition. Without surgery, the mortality rate exceeds 90% at 1 year of age [5]. To date, a few isolated adult interrupted aortic arch patients were reported in the literature, and most of them were treated surgically at the time of diagnosis [7]. The course of patients who declined surgery is not clearly defined in the literature [8]. Benefits of the operation have not been clearly described and complication rates of the surgery have not been established yet.

Conclusions

Herein we described a case of adult isolated interrupted aortic arch treated medically with a multidisciplinary approach. Our patient had good collaterals, which ensured sufficient blood flow to the distal aorta and thus a milder presentation of interrupted aorta, and this was the main cause of the uncomplicated course of the patient at 4-year follow-up. We think isolated interrupted aorta patients who have good collaterals that provide sufficient blood flow to the distal aorta and who declines surgery can be followed safely with medical therapy under team surveillance. In the English literature this is the first case of adult interrupted aorta followed safely for 4 years under medical therapy.