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Case Report

# Interrupted aortic arch in an old woman with aortic stenosis

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**Abstract.** Interrupted aortic arch (IAA) is a rare and usually lethal congenital malformation. Without previous surgical intervention to reach adult age is very rare in patients with complete IAA. The present report describes a 70-year-old hyptertensive women who was incidentally diagnosed to have IAA and aortic stenosis. Aortography showed a complete IAA below the origin of left subclavian artery and gadolinium contrast-enhanced magnetic resonance angiogram (1.5 T scanners) clearly reaffirmed a complete interruption of the arcus aorta, with markedly developed collateral circulation.

Key words: Interrupted aortic arch; hypertension; aortic stenosis

## 1. Introduction

Interrupted aortic arch (IAA) is a rare congenital malformation (1). It is an extreme form of aortic coarctation and is characterized by complete luminal and anatomic discontinuity between ascending and descending aorta (2). Isolated IAA is extremely rare; most of the patients have an associated cardiac anomaly such as ventricular septal defect, patent ductus arteriosus, bicuspid aortic valve and left ventricular outflow tract obstruction (1,2). Prognosis of this anomaly is very poor unless a successful surgery is performed. We describe here a 70-year-old female patient with hypertension and aortic stenosis who had an isolated type A IAA.

## 2. Case report

A 70-year-old woman was referred to our hospital with the complaints of general malaise, exertional dyspnea and headache for two years. She had hypertension for five years. On physical examination, blood pressure was 170/110 mmHg (on left and right arms). Her lower limb blood pressure was measured as 110/70 mmHg and lower extremity pulses were weaker than upper extremity pulses. There was grade 3 systolic murmur on the right second intercostal area. The electrocardiogram showed left ventricular hypertrophy and sinus rhythm. Chest radiography revealed no pathologic abnormality. Transthoracic echocardiography demonstrated severe aortic stenosis (maximum/mean gradient: 90/53 mmHg and aortic valve was tricuspid) associated with concentric left ventricular hypertrophy and preserved left ventricular systolic function. Her liver and renal function tests were normal; ALT: 24 U/L, AST: 17 U/L and BUN: 19 mg/dl, creatinine 1.0 mg/dl. Cardiac catheterization was performed for evaluation of concomittant coronary artery disease. Catheterization through right femoral artery showed that aortic arch was interrupted (Fig. 1).

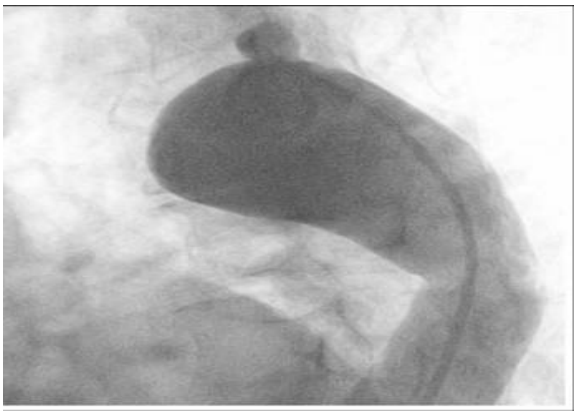

Fig. 1. The complete lack of opacification the aortic arch below the left subclavian artery indicating of complete interruption of the aorta at this level.

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The procedure was continued through right brachial artery. However, coronary anjiography could not be performed through subclavian arteries due to extensive colletaralization between subclavian arteries and aortic arch. Noncontrast enhanced thoracic three dimensional magnetic resonance angiogram (MRA) was performed to clarify the aortic anatomy. The MRA clearly reaffirmed a complete interruption distal to the left subclavian artery and there was an extensive collateralization between major arteries (Figure 2). Multislice computed coronary angiography was not avaliable in our location. The patient was scheduled for multislice computed tomography and elective surgical repair. However, she refused further examination and operation.

Fig. 2. Noncontrast 3-dimensional reconstructed magnetic resonance angiogram showing interrupted aortic arch with extensive collaterals

## 3. Discussion

Interrupted aortic arch is defined as the absence of luminal continuity between the ascending and descending portions of the aorta (3). IAA is an extremely rare congenital malformation of the aorta (3 per million live births) (1). The first classification system, introduced by Celoria and Patton in 1959, is still used almost universally (4). It can be classified into three types, based on the position of the interruption. Type A is distal to the left subclavian artery; type B is between the left common carotid artery and the left subclavian artery and type C is between the brachiocephalic artery and the left common carotid artery (4). The most common type is B (53%), followed by A (43%) and C (4%) (2).

Two-dimensional echocardiography plays an important role in the delineation of IAA. However, echocardiography may be suboptimal for visualizing the arch vessels and distinction between an IAA and aortic coarctation with a hypoplastic aortic arch may be difficult (5). In the present case, we could not observe the IAA with transtoracic echocardiography. We detected the IAA during cardiac catheterization, which was performed to evaluate coronary anatomy since an operation was planned for severe degenerative aortic stenosis. Cardiac catheterization is the most widely used technique for accurate diagnosis of IAA. But cardiac catheterization is an invasive method and it has several limitations. Most important limitation is the inability of the cannulation of the femoral and brachial-axillary arteries. Thoracic 3D MRA is a reliable noninvasive diagnostic modality for the diagnosis of aortic coarctation, aortic arch anomalies and for visualization of collateral vessels (6). The prognosis for untreated infants is extremely poor and survival beyond infancy is uncommon. To the best of our knowledge, the medical literature contains only 21 cases of isolated IAA in adults and as far as we know, our case is the oldest patient with IAA. As in our case, exstensive collateralization must be present to maintain flow and enable survival. In these few documented cases in adults, the presentation is very different (ranging from a lack of symptoms, limb swelling to differential blood pressures in all extremities (1). It is interesting that our case had easily tolerated six pregnancies without symptoms. She had general malaise, exertional dyspnea and headache only for two years, most likely due to severe aortic stenosis.

We can conclude that, patients with complete IAA may survive up to late adulthood without any serious medical problem thanks to extensive collateralization.

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