

Undiagnosed interrupted aortic arch in a 59-year-old male patient with severe aortic valve stenosis: A case report and literature review

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Case Report

Abstract

BACKGROUND: Interrupted aortic arch (IAA) is defined by a lack of the luminal continuity between the ascending and descending thoracic aorta. It is a rare, severe congenital heart defect which without surgery is associated with high mortality in the neonatal period. The aims of this study were to present a case with IAA who was alive until the age of 59 years without any surgical intervention and to review the literatures that have presented IAA cases.

CASE REPORT: The patient was admitted with respiratory distress and pulmonary edema. Echocardiography showed the severe stenosis in aortic valve and severe left ventricular dysfunction. Cardiac catheterization and angiography confirmed interrupted aorta (type A). The descending thoracic aorta was supplied by extensive collateral vessels from the vertebrobasilar system down to the posterior chest wall and the spine. Surgical correction including coronary artery bypass graft and aortic valve replacement and repair of interruption of the aorta was performed. Three weeks later the patient was died due to uncontrollable gastrointestinal bleeding and hospital acquired pneumonia. We described diagnosis and management of our case.

CONCLUSION: This case was very interesting for us, because the patient had not been diagnosed until the recent presentation. Similar cases with this diagnosis do not reach adulthood, but our patient was alive up to 59 years of age.

Keywords: Interrupted, Aorta, Aortic Valve Stenosis, Thoracic Aorta, Aortic Arch

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Introduction

Interrupted aortic arch (IAA) is a rare, severe congenital heart defect defined as a complete loss of luminal and anatomic continuity between ascending and descending aorta,¹ representing approximately 1% of congenital heart disease.² It usually occurs in association with the nonrestrictive ventricular septal defect and ductus arteriosus or, less commonly, with a large aortopulmonary window or truncus arteriosus.³ In the presence of two ventricles, varying degrees of left ventricular (LV) outflow tract

obstruction is often observed.^{4,5} It occurs in three per million live births.^{6,7} IAA has been classified into three types (A, B, and C) based on the site of the aortic interruption. In the type A, interrupted left aortic arch, the arch interruption occurs distally to the origin of the left subclavian artery. In type B, interrupted left aortic arch, the interruption occurs distal to the origin of the left common carotid artery. In the type C, interrupted left aortic arch, the interruption occurs proximally to the origin of the left common carotid artery. Type B interruption

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accounts for about two-third of cases, type A occurs in about one-third of cases, and type C is presented in less than 1% of cases.³

The IAA is a congenital cardiopathy which has devastating consequences, with a 75% mortality rate at 10 days and 90% at 12 months of life.⁶ In infants, its' clinical presentation often involves severe congestive heart failure and if left untreated, most affected infants die within some days. Lobato et al., in their study, reported that few cases were with IAA, which most of them need surgical replacement.⁸

In our case report, we want to introduce a 59-year-old man with undiagnosed interrupted aorta and how we managed him live up to 59 years without further surgical intervention.

Case Report

A 59-year-old man was presented in our hospital because of respiratory distress since 3 days before admission, which had been gradually severe. He was admitted with impression of pulmonary edema. He had a history of uncontrolled diabetes mellitus, systemic hypertension and hyperlipidemia.

On physical examination, he was blind, with blood pressure of 210/140 mmHg, pulse rate about 150 beat/min and respiratory rate about 40 cycle/min. The pulses were equal in upper limbs. Both femoral pulses were equal but weak. There were diffuse moist rhales in both lungs and cardiac examination systolic ejection sound and murmur was audible in the aortic area and with less severity in apex and lower left sternal border.

Electrocardiography showed sinus tachycardia with complete left bundle branch block. After several hours of aggressive medical treatment, the patient's condition became relatively stable.

Echocardiography showed sever aortic valve stenosis (mean pressure gradient = 60 mmHg) and sever LV systolic dysfunction (LV ejection fraction = 25%).

After initial stabilization with conservative treatment, coronary angiography was done and revealed three vessels coronary artery disease.

Cardiac catheterization from right femoral artery showed occlusion of the distal thoracic aorta to the left subclavian artery and angiography from right brachial artery proved interrupted aorta (type A). We were not able to pass through aortic valve to the left ventricle because of valvular stenosis, and it seems that left internal mammary artery plays role in collateralization but we did not engage through it (Figure 1).

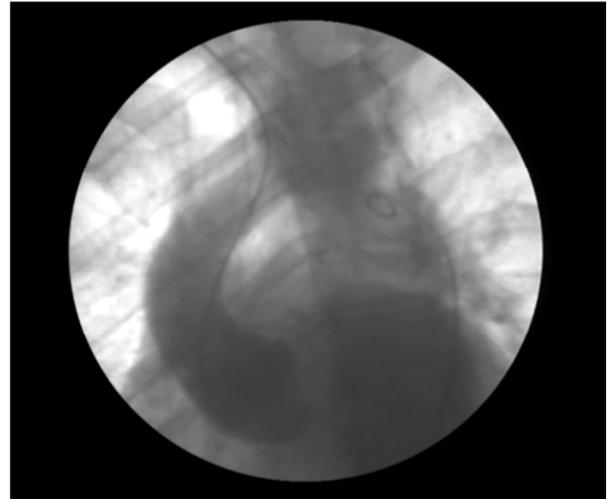


Figure 1. Angiography from right brachial artery proved interrupted aorta

The descending thoracic aorta was supplied by extensive collateral vessels from the vertebrobasilar system down to the posterior chest wall and the spine (Figure 2). Successful surgical correction including coronary artery bypass graft and aortic valve replacement and repair of interruption of the aorta was performed without any complication. Three weeks later the patient died due to gastrointestinal (GI) bleeding which was not controllable by aggressive treatment and hospital acquired pneumonia.

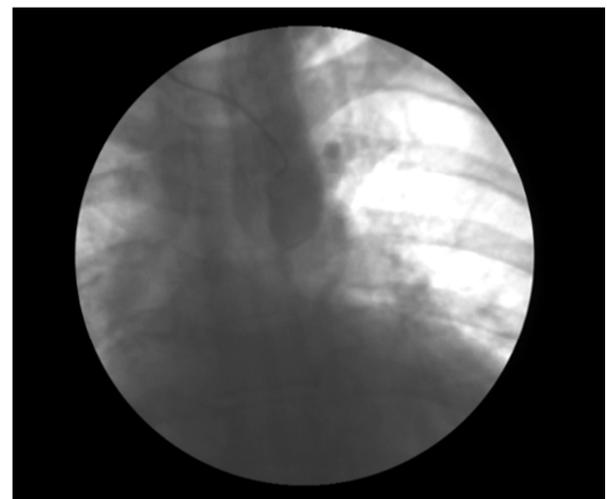


Figure 2. The descending thoracic aorta was supplied by extensive collateral vessels

Discussion

The IAA is a rare, severe congenital heart defect, which without surgery is associated with high mortality in the neonatal period;⁸ but our case until age 59 without any surgery intervention was alive.

This disease displays the absence of communication between the two segments of the thoracic aorta and, consequently, of the blood flow; thus, most cases are expected to be fatal. In our case, cardiac catheterization showed occlusion of the distal thoracic aorta to the left subclavian artery (type A).¹

The common characteristic among the survivors is the presence of an extensive collateral network, which is necessary for the maintenance of the distal flow and the consequent organ viability.⁶

In our case, the descending thoracic aorta was supplied by extensive collateral vessels from the vertebrobasilar system down to the posterior chest wall and the spine.

In this case, surgical correction was performed without any complication but the main reason for death of the patient was uncontrollable GI bleeding and hospital acquired pneumonia.

Conclusion

This case was very interesting for us, because the patient had not been diagnosed up to a recent presentation. Similar cases with this diagnosis do not reach adulthood, but our patient was alive up to his 60th decade.

Conflict of Interests

Authors have no conflict of interests.

References

1. Kleinrok A, Zarembo-Flis E, Smyk T. Interrupted aortic arch in an adult female. *Echocardiography* 2010; 27(7): E70-E72.
2. Kosucu P, Kosucu M, Dinc H, Korkmaz L.

Interrupted aortic arch in a adult: diagnosis with MSCT. *Int J Cardiovasc Imaging* 2006; 22(5): 735-9.

3. Chin AJ. Interrupted aortic arch [Online]. [cited 2011]; Available from: URL: <http://emedicine.medscape.com/article/896979-overview>
4. Vukomanovic V, Stajevic M, Prijic S, Bjelakovic B. Interrupted aortic arch and aortopulmonary window associated with complete atrioventricular septal defect. *Indian Pediatr* 2012; 49(2): 147-9.
5. Shinkawa T, Jaquiss RD, Imamura M. Single institutional experience of interrupted aortic arch repair over 28 years. *Interact Cardiovasc Thorac Surg* 2012; 14(5): 551-5.
6. Canova CR, Carrel T, Dubach P, Turina M, Reinhart WH. Interrupted aortic arch: fortuitous diagnosis in a 72-year-old female patient with severe aortic insufficiency. *Schweiz Med Wochenschr* 1995; 125(1-2): 26-30.
7. Bayraktutan U, Kantarci M, Ceviz N, Yuce I, Ogul H, Sagsoz ME, et al. Interrupted aortic arch associated with AP window and complex cardiac anomalies: multi detector computed tomography findings. *The Eurasian Journal of Medicine* 2012; 45: 62-4.
8. Lobato RF, Saliba LA, Ferreiro CR, Bacal F. Interrupted aortic arch with cardiac heart failure in young adult. *Arq Bras Cardiol* 2008; 91(1): e4-e6.

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