Arch Iran Med. December 2017;20(12):756-759

http www.aimjournal.ir

ARCHIVES OF IRANIAN MEDICINE

Open Access

Case Report

Endovascular Repair of Interrupted Aortic Arch: Approach with Hope for Fewer Complications

Ata Firouzi, MD[1]; Bahram Mohebbi, MD[2*]; Ali Shafiei, MD[3]

[1]Cardiovascular Intervention Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

[2]Cardiovascular Intervention Research Center, Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

[3]Rajaie Cardiovascular Medical and Research Center, Iran University of Medical Sciences, Tehran, Iran

Abstract

Interrupted aortic arch (IAA) is a rare congenital malformation defined as complete discontinuity between ascending and descending parts of aorta. We present a case of IAA, which was referred to us due to dilatation of proximal and mid parts of his thoracic aorta accompanied by narrowing of aorta proximal to the branching of the left subclavian artery. Further evaluation revealed interruption of aorta at the proximal part of descending thoracic aorta by a transverse septum along with several collateral formations. In general, the standard treatment of IAA is open surgical repair. Endovascular repair of IAA is an alternative approach for IAA, which is applied when two distinct parts of aorta are too close to each other. Here, we present a new approach of endovascular transcatheter repair of IAA with implantation of a self-expandable stent that we believe has fewer complications.

Keywords: Angioplasty, Endovascular repair, Interrupted aortic arch, Self-expandable stent

Cite this article as: Firouzi A, Mohebbi B, Shafiei A. Endovascular repair of interrupted aortic arch: approach with hope for fewer complications. Arch Iran Med. 2017;20(12):756–759.

Received: January 18, 2017, Accepted: December 9, 2017, ePublished: December 23, 2017

Introduction

Interrupted aortic arch (IAA) is defined as complete lack of continuity between ascending and descending parts of aorta.[1-5] It is a rare congenital malformation seen in 3 cases per 1 000 000 live births.[2,4] However, it is very rare in adults.[2,3] It was described for the first time by Steidele in 1778.[2] IAA is classified by Celoria and Patton in 3 types: type A in which discontinuity is located distal to the left subclavian artery; type B between the left common carotid artery and the left subclavian artery; and type C between the brachiocephalic artery and the left common carotid artery.[6]

Case Report

The patient was a 46 years old man who referred to Rajaie Cardiovascular Medical and Research Center, Tehran, Iran for repair of aneurysm of ascending aorta. He was referred to our center from the city of Kashan, Iran. CT-angiography performed for him in Kashan had revealed dilatation of proximal and mid parts of ascending aorta with the length of 57 mm and width of 54 mm accompanied by narrowing of aorta proximal to the branching of left subclavian artery. He did not have any history of hypertension, dyslipoproteinemia, diabetes mellitus, or cigarette smoking. The only findings in his physical examination were a bounding pulse of carotid and an early diastolic murmur. Transesophageal echocardiography was performed for him which showed severe left ventricular enlargement and systolic dysfunction with a left ventricle ejection fraction (LVEF) of only 25%, hypertrabeculated left ventricular apical segment with severe global hypokinesia, a grade II left ventricle diastolic dysfunction, severe right ventricular enlargement with moderate systolic dysfunction, mild to moderate biatrial enlargement, mildly thickened mitral valve with up to moderate functional mitral regurgitation (MR), dilated tricuspid valve (TV) annulus with the diameter of 46mm resulting in at least moderate eccentric tricuspid regurgitation (TR) (TRG: 52 mm Hg), mildly thickened and tricuspid aortic valve with severe aortic insufficiency (AI), aneurysmally dilated ascending aorta with the width of 54 mm at its tubular portion, resulting in ST junction effacement and severe AI, shelf-like protrusion in proximal descending thoracic aorta at ductus level resulting in severe narrowing (the width was only 3 mm) and significant coarctation as there was no significantly increased trans-stenosis gradient registered due to Doppler malalignment and multiple collaterals, and relative narrowing of abdominal aorta with slow systolic upstroke and antegrade diastolic flow in spectral Doppler

***Corresponding Author:** Bahram Mohebbi, MD; Rajaie Cardiovascular Medical and Research Center, Valiasr Avenue, Postal Code: 19969-11156, Tehran, Iran. Tel: +98-21-23922072, Fax: +98-21-22055594, Email: roodbar@yahoo.com.

study which are consistent with significant coarctation of aorta. The other findings in his TEE were moderate to severe pulmonary artery hypertension (PAH) and no ventricular septal defect (VSD) (Table 1). Spiral CT-angiography of thoracic and abdominal aorta and other organs with special reconstruction views with dynamic contrast medium was performed for the patient which revealed interruption of aorta at the proximal part of descending thoracic aorta by a transverse septum along with several collateral formations (Figure 1).

Based on clinical presentation and transesophageal and spiral CT-angiographic findings, it was revealed that the patient was a case of IAA and the two interrupted parts of his aorta were separated from each other only by a transverse septum. Therefore, it was decided to manage the patient by a endovascular transcatheter repair in which a self-expandable stent was applied instead of balloon-expandable stent, because it was believed that this approach comes with fewer complications and better adaptation with the vessel wall. First, femoral artery access was implanted but wire passage from IAA was not possible via femoral artery access, so wiring was performed via transradial access, then sequential predilation was done with Maverick balloon 2 mm × 9 mm, Filao NC 4mm × 12 mm, and Wanda balloon 5 mm × 40 mm, followed by stenting with sinus-XL stent 22 mm × 61 mm with good final results without any complication.

Additional angiographic findings included normal epicardial coronary arteries, severe AI, annuloaortic ectasia, aneurysmally dilated ascending aorta, and severe LV systolic dysfunction (LVEF: 25%).

The patient was transferred to the CCU for one day and then transferred to the post CCU ward. Two days later, he was discharged from our center in good clinical condition.

The patient also had severe aortic insufficiency that needed to be treated by Bental procedure; therefore, the author decided to perform surgical treatment of the patient's severe AI after rehabilitation of endovascular treatment of his IAA.

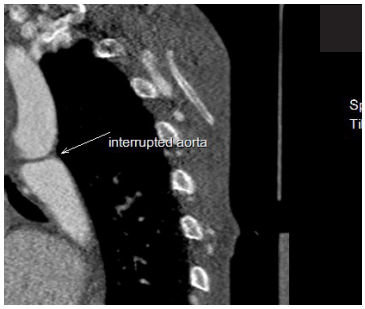

Figure 1. Spiral CT-Angiography of Thoracic and Abdominal Aorta.

Table 1. Transesophageal Echocardiography Findings

1	Systolic dysfunction (LVEF: 25%)
2	Severe global hypokinesia
3	LV diastolic dysfunction (grade II)
4	Severe right ventricular enlargement with moderate systolic dysfunction
5	Mild to moderate biatrial enlargement
6	Severe AI
7	Aneurysmally dilated ascending aorta
8	Severe narrowing of proximal descending thoracic aorta
9	Multiple collateral vessels at the site of severe narrowing
10	Slow systolic upstroke in spectral Doppler study
11	Antegrade diastolic flow in spectral Doppler study

Discussion

IAA is a rare congenital malformation which occurs in 3/1 000 000 live births.[1,2] This malformation is defined as complete discontinuity of ascending and descending parts of aorta.[1-5] According to Celoria and Patton, based on the site of disintegration, IAA is classified in 3 distinct types: type A in which discontinuity is located distal to the left subclavian artery; type B in which discontinuity is located between the left common carotid artery and the left subclavian artery; and type C in which discontinuity is located between the brachiocephalic artery and the left common carotid artery.[6] Type B is the most common type (53%), followed by type A (43%), and finally type C (4%).[6] IAA is usually accompanied by other cardiovascular anomalies such as arterial septal defect (ASD), VSD, patent ductus arteriosus (PDA), bicuspid aortic valve, left ventricular outflow tract obstruction, double outlet right ventricle, aortopulmonary window, truncus arteriosus, and DiGeorge syndrome.[1,4,5]

The most common presentation of IAA in neonates is severe congestive heart failure, and without treatment, 90% of patients die at a median age of 4 days.[2,3]

In adults, IAA is quite rare and only a few documented cases are presented in medical literature.[2,3] However, according to Mirat and colleagues, the incidence of IAA may be different in some adult patients due to lack of development of symptoms or signs even for several decades.[7] The existence of developed collaterals is essential for reaching adulthood[8]; however, these collateral vessels may experience atrophy, atherosclerosis or spontaneous rupture, and subsequently secondary complications.[3] In adults, the clinical presentation of IAA has a spectrum from asymptomatic patients to differential blood pressure in the extremities or even isolated hypertension[2,3,7]

Comprehensive history and careful physical

examination are the first step for diagnosis of IAA, especially to detect blood pressure in upper and lower extremities at the same time in patients with isolated hypertension especially in young ones: the difference between the blood pressure of right and left upper extremities or upper and lower extremities is an important clue for diagnosis of coarctation of aorta or IAA. However, to confirm the diagnosis of IAA, some imaging studies are needed, including echocardiography, CT-angiography, MRI, and MR-angiography.[8]

In general, the standard treatment for IAA is open surgical treatment, and today the most common approach is either direct end to end anastomosis of two parts of aorta or patch aortoplasty of these two separated segments, depending on the distance between the interrupted parts. Surgical repair has some complications, most importantly paraplegia in approximately 0.5%–1% of cases, paradoxical hypertension, restenosis, and coarctation or aneurysm formation at the site of surgical repair.[9,10] Endovascular repair of IAA is an alternative approach for IAA, which is applied when the two interrupted parts of aorta are too close to each other or when these two parts are separated by only a transverse septum as in our case. It is thought that in some cases, a potential stalk passes across the transverse septum; so, in endovascular approach, it is tried to pass the potential stalk by an inflatable balloon and inflate it followed by sequential larger balloon inflation to make a larger tunnel in the transverse septum for stent passage. The patients are treated either by balloon angioplasty alone or along with stenting. However, in general, it is observed that most interventionists prefer stent implantation to balloon angioplasty in situations where endovascular repair by each method is possible.[11] The superiority of stent implantation to simple balloon angioplasty is due to the fact that balloon dilatation may cause some problems such as tears of intima and media; therefore, despite initial increase of vessel diameter, it may cause scar tissue formation which results in restenosis and aneurysmal formation.[10,11] However, stent implantation causes radial support of vessel wall and the apposition of the torn intima to the media.[10,11] Endovascular repair by stent implantation also has some complications including rupture of aorta which is the most serious complication resulting in severe hemothorax and even death, aneurysmal formation, stent migration, and restenosis. However, the rate of restenosis after stent implantation is lower than balloon angioplasty alone.[11] In addition, implantation of self-expandable stents has some advantages over balloon expandable stents; for example, balloon expandable stents have higher rate of stent migration and/or embolization, and higher rate of making trauma during implantation of stent, whereas self-expandable stents have easier deployment, lack of shortening on expansion, and better adaptation to the aortic wall.[12]

Our case was an IAA patient in whom the two parts of aorta were separated from each other by only an interrupted transverse septum of aorta; therefore, he was a suitable case for endovascular repair of IAA. The authors believe that sometimes there is a stalk inside the transverse septum so it is possible to remove the septum by passing predilation balloon through this potential stalk, then implant a self-expandable stent. The authors believe that this approach is less complicated and better tolerated by the patient. On the other hand, surgical repair of the patient's other lesions including aneurysmally dilated ascending aorta associated with severe AI needs a surgical approach (e.g. midsternotomy) which is different from surgical approach to IAA (e.g. thoracotomy) and this makes surgical correction of all lesions of the patient very complex. So, in this patient endovascular stenting of IAA obviated surgical repair of IAA and relatively simplified the surgical approach to patient for repair of reaming lesions including aneurysmally dilated ascending aorta associated with severe AI which will be performed later.

The authors reviewed the literature but did not find any similar approach; the authors do not claim but believe that this approach has some novelty.

Authors' Contribution

AF and BM participated in conducting the practice and designing case report. AS wrote the draft of manuscript. All authors revised subsequent drafts of the paper. BM prepared this manuscript for publication.

Conflict of Interest Disclosures

The authors have no conflicts of interest.

Ethical Statement

Written informed consent was obtained from the patient for publishing this case report.

References

1. Hsieh YC, Wu TJ, Wang KY, Liang KW, Lin WW, Chen YT, et al. An adult with aortic arch interruption associated with sinus venosus atrial septal defect and partial anomalous pulmonary venous connection. J Chin Med Assoc. 2007;70(1):30-2. doi: 10.1016/s1726-4901(09)70297-7.
2. Messner G, Reul GJ, Flamm SD, Gregoric ID, Opfermann UT. Interrupted aortic arch in an adult single-stage extra-anatomic repair. Tex Heart Inst J. 2002;29(2):118-21.
3. Sai Krishna C, Bhan A, Sharma S, Kiran U, Venugopal P. Interruption of aortic arch in adults: surgical experience with extra-anatomic bypass. Tex Heart Inst J. 2005;32(2):147-50.
4. Bugan B, Iyisoy A, Celik M, Kucuk U, Boz U, Celik T. Isolated type a interrupted aortic arch: in an asymptomatic 19-year-old man. Tex Heart Inst J. 2011;38(5):559-61.
5. Mishra PK. Management strategies for interrupted aortic arch with associated anomalies. Eur J Cardiothorac Surg. 2009;35(4):569-76. doi: 10.1016/j.ejcts.2008.12.044.
6. Celoria GC, Patton RB. Congenital absence of the aortic

arch. Am Heart J. 1959;58(3):407-13. doi: 10.1016/0002-8703(59)90157-7.
7. Mirat J, Galic E, Coric V, Rajsman G, Vrbanic L, Razov Radas M, et al. Secondary hypertension due to isolated interrupted aortic arch in a 60-year-old person--one-year follow up. Coll Antropol. 2010;34 Suppl 1:307-9.
8. Dillman JR, Yarram SG, D'Amico AR, Hernandez RJ. Interrupted aortic arch: spectrum of MRI findings. AJR Am J Roentgenol. 2008;190(6):1467-74. doi: 10.2214/ajr.07.3408.
9. Mullen MJ. Coarctation of the aorta in adults: do we need surgeons? Heart. 2003;89(1):3-5. doi: 10.1136/heart.89.1.3.
10. Tzifa A, Ewert P, Brzezinska-Rajszys G, Peters B, Zubrzycka M, Rosenthal E, et al. Covered Cheatham-platinum stents for aortic coarctation: early and intermediate-term results. J Am Coll Cardiol. 2006;47(7):1457-63. doi: 10.1016/j.jacc.2005.11.061.
11. Godart F. Intravascular stenting for the treatment of coarctation of the aorta in adolescent and adult patients. Arch Cardiovasc Dis. 2011;104(12):627-35. doi: 10.1016/j.acvd.2011.08.005.
12. Haji-Zeinali AM, Ghasemi M. Coarctoplasty with self-expandable stent implantation for treatment of coarctation of aorta in adults. Arch Iran Med. 2006;9(4):348-53.