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

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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

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 an 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 4 mm × 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.

**Discussion**

IAA is a rare congenital malformation which occurs in 3/1,000,000 live births. This malformation is defined as complete discontinuity of ascending and descending parts of aorta. 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. Type B is the most common type (53%), followed by type A (43%), and finally type C (4%). 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.

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. In adults, IAA is quite rare and only a few documented cases are presented in medical literature. 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. The existence of developed collaterals is essential for reaching adulthood; however, these collateral vessels may experience atrophy, atherosclerosis or spontaneous rupture, and subsequently secondary complications. In adults, the clinical presentation of IAA has a spectrum from asymptomatic patients to differential blood pressure in the extremities or even isolated hypertension.

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. 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. 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. 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. However, stent implantation causes radial support of vessel wall and the apposition of the torn intima to the media. 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. 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.

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.

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