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Case Report

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Treatment of Interrupted Aorta in Adult Patients; a Challenge Both in Surgery and Transcatheter Intervention

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Abstract

Interrupted aortic arch (IAA) is a rare congenital cardiovascular condition. We present two cases with completely obstructed descending aortas, just distal to isthmus and that were treated with stent implantation and surgery. The decision depends on the invasive cardiologist's and experience of the surgeon and on the patient's condition.

Key words: Interrupted aortic arch, surgery, transcatheter intervention

Introduction

IAA is defined as the complete loss of luminal and anatomical continuity between the ascending and descending thoracic aorta (1). In most patients, IAA is fatal within the first week of life (2). Although patients with isolated IAA rarely reach adult age without proper surgical intervention, some may live until adulthood due to rich collateral circulation.

The main treatment for IAA is the reconstruction of aortic continuity, usually by surgical or by a percutaneous approach in selected cases (3). IAA is a rare condition that remains a surgical challenge, particularly when associated with other complex cardiac abnormalities, or when there is noncardiac comorbidity (4). We present two cases with obstructed descending aorta just distal to the subclavian artery were treated using the covered stent and surgery.

Case-1

A 44-year-old female house builder presented to our outpatients service because of a history of effort intolerance, general malaise and cough that began two years earlier. Physical examination revealed grade 4 systolic murmur in the auscultation position of the aortic valve. Transthoracic and transesophageal echocardiography showed aortic stenosis and left atrial myxoma. Aortic valve was found to be calcific with severe gradient (max 94, mean 56 mmHg). The patient was scheduled for cardiac catheterization to perform coronary angiography and aortography. Cardiac catheterization was performed by percutaneous femoral artery approach. Since we could not advance any catheter and guide wires retrogradely from the descending to the ascending aorta, brachial artery puncture was performed and a catheter was placed antegradely. Subsequently, aortography from the ascending aorta through the right brachial artery showed normal coronary arteries and complete interruption of the aortic arch approximately distal to the origin of the left subclavian artery (Figure-1). A computed tomography showed an obstruction of the aortic arch distal to the origin of the left subclavian artery and markedly ascending aortic arch. She was performed a single-stage operation involving mechanical aortic valve replacement and left atrial myxomal resection with concomitant anatomic bypass from the ascending to the descending aorta with a Dacron tube graft. The patient made a full recovery.

Case-2

A 28-year-old female presented with weakness of her legs, headache and hypertension that began six months earlier. She had suffered from effort intolerance since childhood. On admission arterial blood pressure was 170/100 mmHg in both arms. Physical examination revealed regular heart rate, grade 3 systolic murmur in the auscultation position of the aortic valve. Bilateral femoral, popliteal pulses were extremely weak and dorsalis pedis pulses were impalpable on physical examination.

Transthoracic echocardiography showed mild left ventricular hypertrophy. Echocardiographic examination could not reveal exactly whether the patients had coarctation or interruption. Cardiac catheterization was performed by percutaneous femoral artery approach. Multiple attempts to cross the interrupted point were unsuccessful. This times cardiac catheterization was performed by percutaneous left brachial artery approach, revealing aneurysm of ascending aorta (Figure-2). Aortography revealed complete interruption of the aortic arch just distal to the origin of the left subclavian artery and there was a small PDA (Figure-3).

Both a cardiothoracic surgeon and adult cardiology interventionalist were consulted to determine the most appropriate therapeutic option.

The atretic segment was retrogradely perforated with the super stiff end of the nitinol guidewire. In addition to the previously placed nitinol guidewire, a coronary guidewire with a soft tip was fed through the same catheter and advanced along the descending aorta into the ascending aorta. The first balloon dilatation was performed with a 3x20 mm coronary balloon. Than dilatation were repeated with a 4,5x20 mm balloon. Angiographies were repeated after post-dilatation and a 22 mm CP stent was implanted (Figure-4). At the end of the procedure control angiograms and pullback measurements were performed. The stent was in good position.

Figure-1 A**:** Retrograde contrast injection reveals no contrast in the transverse aorta. **B**: Anterograde contrast injection reveals the absence of contrast crossing into the descending aorta.

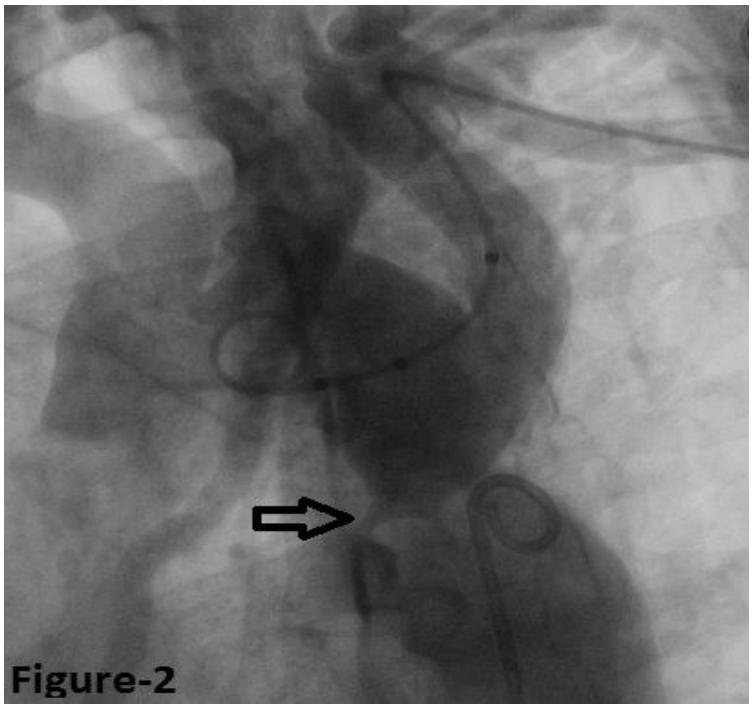

Figure-2: Arch aortogram, showing atresia of the aortic isthmus and small PDA (black arrow).

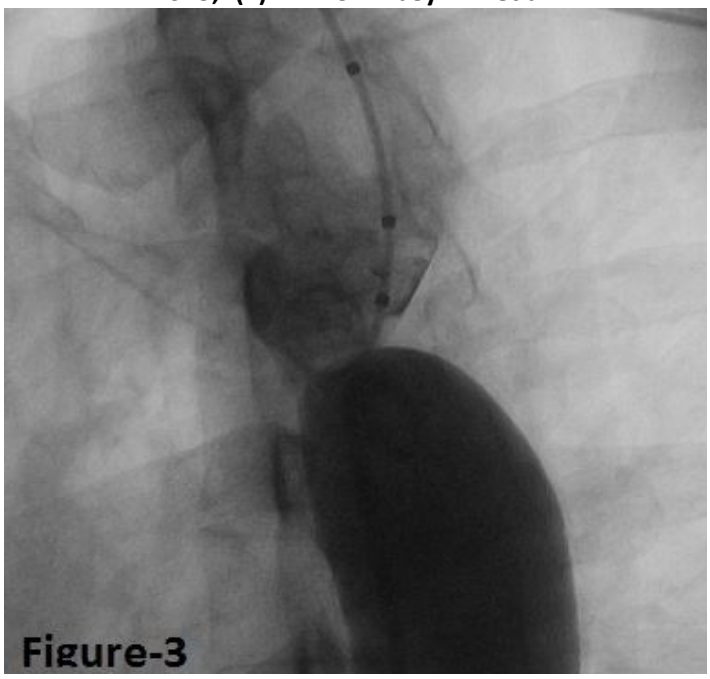

Figure-3: Simultaneous aortography above and below showing atresia of aortic isthmus.

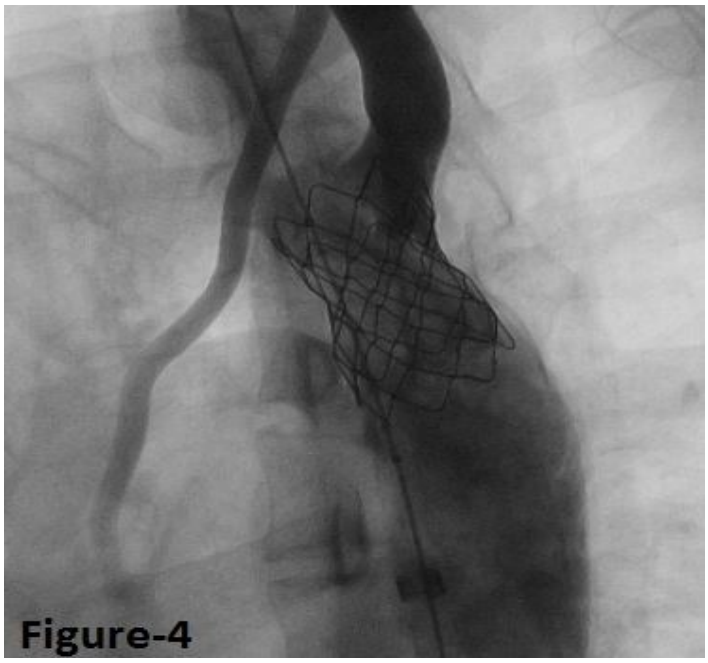

Figure-4: Aortography after stent implantation.

Discussion

Most patients with IAA are detected at a very young age because these patients are critically ill during the neonatal period and need urgent surgical repair (5). IAA may be treated surgically or transcatheter approach. The choice of the therapeutic option should be based on patient wish and preference (6). In the presence of complete luminal obstruction of the aorta, however, surgical repair has been the undisputed and universally recommended therapy (7). Conventional indications for stenting include unfavorable anatomy for balloon angioplasty, such as tubular coarctation, long stenosed aortic segment, hypoplasia of the isthmus and the lack of the effect of balloon angioplasty (8). However, transcatheter approaches are now used more commonly and in some centers they have become a first line treatment in all adult patients, and in some skilled centers stents are used as a palliative treatment even in small children (6, 8). Studies suggests that the advantages of stents compared to balloon angioplasty include significantly lower residual pressure gradient and lower rates of restenosis and direct aortic wall damage (9).

The decision of transcatheter approach or surgery depends on the surgeon and invasive cardiologist's experience and on the patient's overall condition. In first patient, the interrupted aortic arch was accompanied by aortic valve stenosis, calcific aortic valve and left atrial myxoma that necessitated surgical intervention to prevent further deterioration of cardiac function. In second patient, the interrupted aortic arch was accompanied a small PDA and were performed balloon angioplasty and stent implantation successfully.

In conclusion, complete aortic atresia has traditionally been thought to require surgical repair. However, transcatheter treatment is a feasible strategy in selected patients of local atresia of

the aortic isthmus in skilled centers. With selected cases, appropriate anatomy and stenting may be safely and successfully performed. So the treatment of IAA should be individualized.

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