Case report

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Covered stent implantation in an adult with aortic isthmus atresia

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Objective – To present the successful stenting of an aortic isthmic atresia in adult. **Case report** – Aortic isthmic interruption/atresia diagnosed in an adult is a rare congenital defect. Survival from childhood into adulthood is possible if sufficient collaterals have developed during childhood. Using numerous technical and diagnostic skills, the complete luminal continuity between the aortic arch and the descending aorta was established, with stenting of the aortic isthmus without any residual gradient. Only case reports are available regarding this treatment. **Conclusion** – It is possible to treat aortic isthmic interruption/atresia successfully using interventional catheterization, however this treatment may be associated with significant danger of morbidity.

Introduction

Percutaneous stent implantation is the preferred treatment mode in adults with native – or recurrent coarctation of the aorta in many centers (1, 2). Rarely, complete interruption or atresia of the aorta is diagnosed in an adult patient. Survival into adulthood with this malformation is only possible if sufficient collaterals have developed between the upper and lower part of the body (3). Aortic atresia results in complete loss of anatomical and luminal continuity between the ascending and descending aorta, and so far surgery, usually with a tube graft interposition, has been suggested (4, 5). The symptom leading to the correct diagnosis is arterial hypertension in the upper body with diminished or absent femoral arterial pulse, and a significant clinical blood pressure gradient between the upper and lower limbs. Clinically, many patients may be asymptomatic for years, but they experience all the negative side effects of long lasting arterial hypertension. Sometimes aortic atresia is found accidentally during examination for coronary artery disease.

Adequate antihypertensive medical treatment can only be initiated after the anatomical problem has been solved, hence, the atretic segment is either bridged surgically or by interventional means using a catheter. If the interventional catheter approach is employed, the atretic region needs to be crossed to perform stent implantation from the groin. So far, successful catheter interventional treatment in this difficult patient group has only been reported in some case reports and two case series (6-9).

We report the successful stenting of an aortic isthmus atresia in one more adult patient.

Case report

A fifty-eight year old male patient (183cm, 95 kg) presented with arterial hypertension in his right arm (blood pressure 150/80 right arm, 100/50 right leg), NYHA II-III, absent femoral pulses and declining exercise tolerance. He was on medication with valsartan, metroprolol, lercanidipin and hydrochlorothiazide (four antihypertensive drugs). The diagnosis of coarctation with well-established collateralization had been made twenty-two years before, yet the patient did not want to take the risk of invasive therapy. Now a CT scan showed an aortic root dilatation up to 58 mm in diameter, where the aortic valve was bicuspid and mildly stenotic, and the ascending aorta needed to be replaced to avoid aortic rupture. A 3-4 mm gap was present between the distal aortic arch and the descending aorta (Fig. 1).

Fig. 1. *CT-scan showing a bicuspid aortic valve and a dilated aortic root (58 mm). Aortic isthmus atresia with a 3-4 mm gap between the distal aortic arch and the thoracic aorta may be seen. There are multiple collaterals between the upper and lower part of the body.*

The Bentall procedure (aortic root replacement with aortic valve replacement) was thought to be high risk/impossible, without previously addressing the isthmus atresia. Within the "Heart Team" (cardiothoracic surgeon, catheter interventionalist and cardiologist) it was decided to treat the aortic isthmus atresia first in the catheterization laboratory. Under general anesthesia (intubation), the left brachial artery (5F sheath) and the right femoral artery (6F sheath) were cannulated and 5000 units of heparin were given. A 5F pigtail catheter was advanced from the left brachial artery to the distal aortic arch,

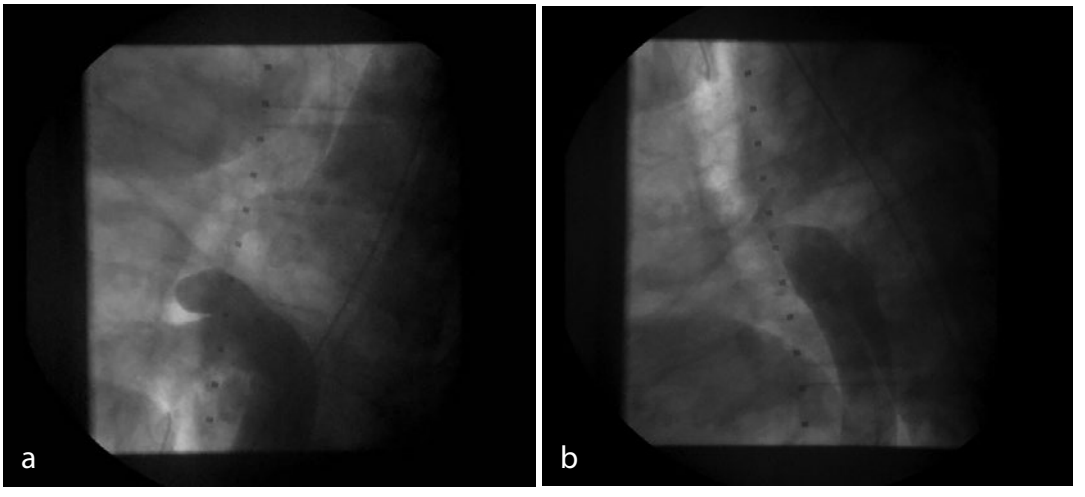

Fig. 2. *Contrast injections into the distal aortic arch via a 5F pigtail catheter advanced from the left brachial artery (a) and into the thoracic aorta (b) through a 6F pigtail catheter introduced through the right femoral artery.*

and a 6F pigtail catheter was placed into the thoracic aorta from the groin.

The peak invasive pressure gradient between the aorta ascendens and descendens was 46 mmHg (AoA 110/53/72; AoD 64/47/53 mmHg, respectively). Angiographies (Fig. 2) showed the aortic isthmus atresia.

It was not possible to cross the atresia from above or below with different guide wires. An 8.5F Swartz™ 45 degree angled braided sheath (St Jude Medical; St Paul MN, USA) was advanced from the right femoral artery close to the atretic segment. The Brockenbrough needle was straightened and advanced through the atretic segment into the distal aortic arch by direct puncture (Fig. 3).

A 0.014' coronary wire was then advanced through the transseptal needle and snared from above with a goose-neck snare 10 mm (Amplatz goose neck EV3, Plymouth, MN, USA). Then the transseptal sheath was advanced into the upper aortic segment (Fig. 4). This sheath was exchanged for a 14F straight long sheath (Cook Europe, Bjaeverskov, Denmark) over a stiff 0.035' guide wire (Amplatz, superstiff, Boston Scientific, Waterston, MA, USA) (Fig. 5).

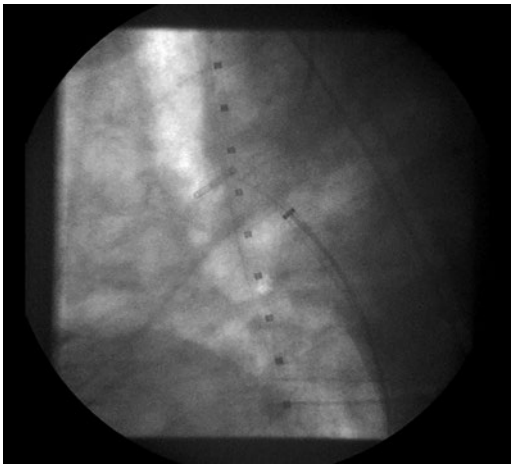

Fig. 3. *A transseptal Brockenbrough needle was straightened and advanced through the 8.5F sheath directly without the introducer shaft. The pigtail 5F serverd as a landmark for the puncture.*

A 45 mm covered Cheatham Platinum stent (NuMed Cornwall, Ontario, Canada) was implanted on an 18 mm BioEnterics Intragastric Balloon (BIB) (NuMed). The stent was not fully expanded (Fig. 6) and left with a waist and residual gradient of 15 mmHg. Three months later, the stent was expanded to 14 mm diameter, resulting in equal pressure in the aorta ascendens and descendens (Fig. 7). The patient underwent the Bentall

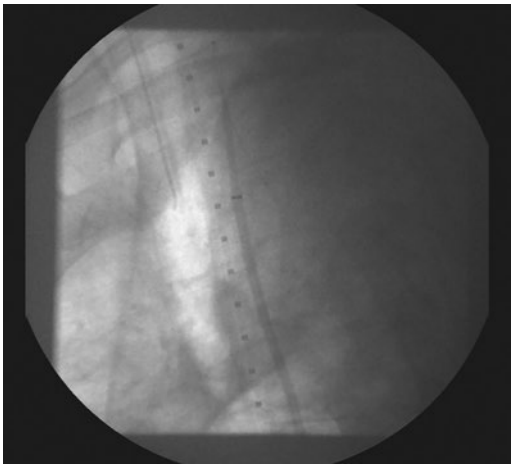

Fig. 4. *A 0.014 coronary wire was pushed through the transseptal needle and snared from above with a 10 mm snare. Then the 8.5F sheath was safely pushed upwards into the distal aortic arch.*

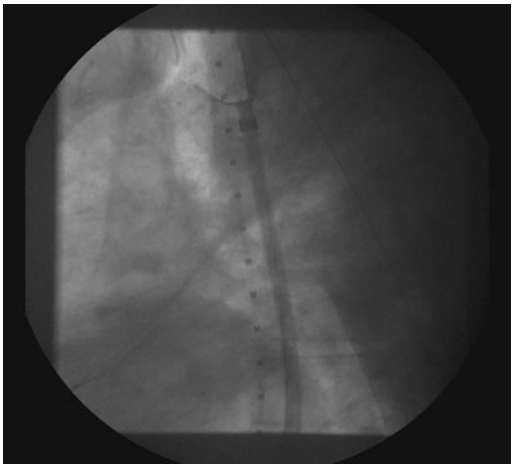

Fig. 5. *The 8.5F sheath was exchanged for a 14F straight long sheath on a 0.035' Amplatz Super Stiff Guidewire.*

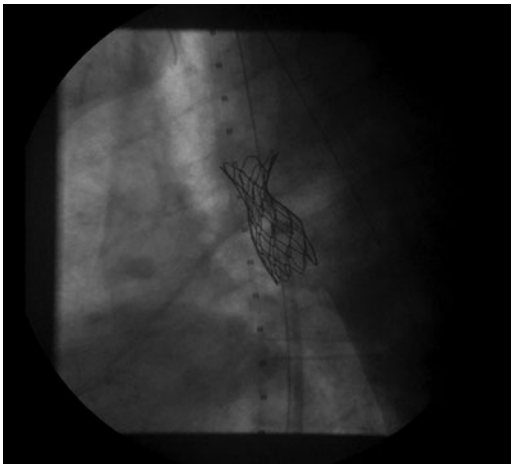

Fig. 6. *A 45 mm covered 8z Cheatham Platinum stent was implanted with an eighteen mm BiB balloon. The stent was not fully dilated. The gradient was reduced from 46 to 15 mmHg.*

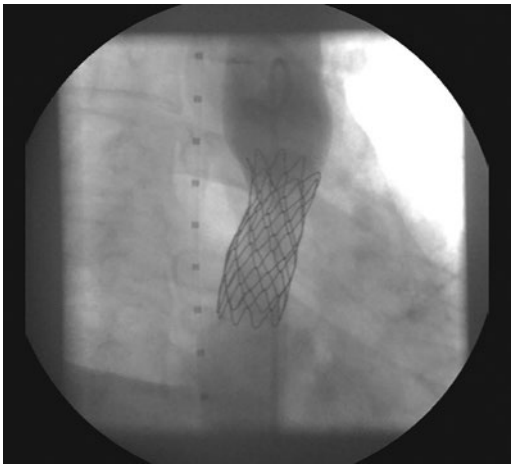

Fig. 7. *Three months later, the stent was dilated to 14 mm diameter. The pressures in the ascending and descending aorta were equal (gradient 0 mmHg).*

procedure and had an uneventful post-operative course. Now, more than six years later, the patient is normotonic (120/70 at the right arm) without a clinical blood pressure gradient to the lower limbs. He is classified as NYHA I, taking only metoprolol and hydrochlorothiazide.

Discussion

We present an adult patient with aortic isthmus atresia and severe arterial hypertension in spite of four antihypertensive drugs, who was treated successfully by catheter interventional covered stent implantation. The gradient between the aorta ascendens – and descendens was abolished, with a sustained good clinical result. The patient is now categorized as NYHA I, normotonic on two antihypertensive drugs.

Aortic isthmic interruption/atresia diagnosed in an adult is a rare congenital defect (10). Until recently, surgical treatment, usually a tube graft interposition, was recommended. However, only a few case reports are available on this treatment (5), which may be associated with significant morbidity: bleeding (collaterals), paraplegia, pleural effusions and laryngeal nerve palsy.

While stent implantation has emerged as the first line treatment for adult native – or recurrent coarctation of the aorta in many centers, only a few case reports (6, 7, 11) and two case series on the treatment of aortic isthmus atresia (8, 9) have been published. The main technical problem in this intervention is to pass the gap between the caudal and cranial aorta safely, without significant aortic bleeding. In our patient we used a modified Brockenbrough needle for the puncture. A pigtail catheter was left in the upper aorta as a landmark. A 0.014' coronary wire was advanced through the needle, which was snared from above. Hence, it was possible to push the sheath up safely over this rail. Others proposed the use of a radiofrequency system (Baylis MedComp Inc, Montreal, Canada) for this step of the intervention (8). Most patients in the largest series so far by Momenah et al. (9) were treated the same way as our patient.

The introduction of balloon expandable covered stents has added safety to the interventional treatment of patients with aortic coarctation, and it allows us to treat patients

with aortic isthmus atresia in the catheterization laboratory (12). We tend to leave the stent diabolo shaped with a central waist at the initial intervention. Later on, the stent is then expanded to its final configuration.

Conclusion

In conclusion, catheter interventional treatment of aortic isthmus atresia in an adult patient is feasible. The safety of this intervention needs to be proven in a larger case series.

Conflict of interest: The authors declare that they have no conflict of interest.

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