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Case Report

Aortic Arch Bare Metal Stenting in a 10-Years-Old Boy with DiGeorge Syndrome Using Overdrive External Pacing

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Abstract

Introduction: In the last decade, percutaneous stent placement has become the standard for treating CoA after childhood. However, stent placement as a treatment for aortic arch hypoplasia has only been described in small series and case reports.

Case Report: A nine-year-old boy with DiGeorge syndrome (BW 50 kg, BL 140 cm, BSA 1.4 m2) was admitted to our hospital due to planned heart catheterisation with aortic arch stenting. In the neonatal period, he underwent complete surgical correction of the IAA type B with perimembranous VSD. During the follow-up period, the progressive narrowing of the aortic arch was observed with gradually increasing systolic pressure gradient (PG) on the echocardiography examination (PG 55 mmHg). The cardiac catheterization was performed under general anaesthesia. The peak-to-peak gradient was estimated at 32 mmHg (AoAsc 128/78 mmHg; AoDsc 96/68 mmHg). Aortography pointed out tubular hypoplasia of the aortic arch between branches (17 mm in length) with localized narrowing right behind the first aortic branch, 6 mm in diameter, and post-stenotic dilatation. The stenosis was resolved using a 22mm long uncovered Cheatham Platinum (CP) stent crimped on a 12 mm Balloon-in-Balloon (BiB) catheter with additional flaring of the distal stent part (Figure 1B and C). During the stent implantation, an overdrive external pacemaker (wire was placed into the right ventricle anterograde across the right femoral vein) was used. Control angiography relieved satisfying anterograde flow across the aorta and aortic branches. The peak-to-peak gradient after stenting was 10 mmHg.

Conclusion: Although technically challenging, carefully analyzing the arch and vessel morphology allows the dilation of the transverse aortic arch hypoplasia and stenosis using a bare metal stent with good angiographic and hemodynamic results and without complications.

Keywords: *Aortic arch stenting, Uncovered CP stent, Interrupted aortic arch, DiGeorge Syndrome*

Introduction

Steidele described an interrupted aortic arch (IAA) in 1778. It can be considered the most severe form of aortic coarctation (CoA), which affects approximately 1.5% of congenital heart disease (CHD), while almost 50% of patients with IAA have DiGeorge syndrome. In these patients, the posterior malalignment of the conal septum produces a ventricular septal defect (VSD) as the most common associated lesion (in 80-90% of all cases) (1,2). Most reports highlight type B morphology as the most common form (52—90%). After one stage of IAA surgery with VSD, overall, 5-year survival is 60–80%, and 5-year freedom from reintervention for arch obstruction is 60–80% (3). Song et al., found that 24/152 patients developed aortic arch stenosis after IAA repair, and one of the independent risk factors was anastomosis with autologous pericardium to the anterior aortic wall (3). On the other hand, in the study conducted by Andrianova et al., 19% of patients required reintervention after surgical correction of IAA and independent risk factors for reintervention were complex IAA, use of an interposition graft and direct anastomosis with a patch (4). Although poor outcomes have been reported for balloon dilation of the transverse arch alone, stenting of the transverse arch has been described in a small series but with good initial results.

We presented a 10-year-old boy with DiGeorge syndrome whose transverse aortic arch stenting was performed after the complete surgical correction of IAA with VSD.

Case Report

A nine-year-old boy (BW 50 kg, BL 140 cm, BSA 1.4 m^2) was admitted to our hospital due to planned heart catheterisation with aortic arch stenting. In the neonatal period, he underwent complete surgical correction of the IAA type B with perimembranous VSD - direct end-to-side anastomosis with VSD closure with a pericardial patch. The postoperative course was complicated with hypocalcemia. The genetic analysis revealed Di-George syndrome. During the follow-up period, the progressive narrowing of the aortic arch was observed with gradually increasing systolic pressure gradient (PG) on the echocardiography examination (PG 55 mmHg). The noninvasive pressure on the right hand was 30 mmHg higher than the pressure on the right leg. A joint cardiology/cardiothoracic surgery (JCC) meeting decided to perform thoracic endovascular aortic repair (TEVAR).

The cardiac catheterization was performed under general anaesthesia. The pigtail catheter was retrogradely placed in the ascending aorta across the right femoral artery. Heparin 100 U/kg was given once vascular access was obtained by a femoral artery puncture. The peak-to-peak gradient was estimated at 32 mmHg (AoAsc 128/78 mmHg; AoDsc 96/68 mmHg). The aortography was performed in the left lateral (LL) and left anterior oblique (LAO) 50 positions. Aortography pointed out two aortic arch branches – the first was a common origin of truncus brachiocephalic and the left carotid artery, and the second was the left subclavian artery. Between those two branches, tubular hypoplasia of the aortic arch (17 mm in length) with localized narrowing right behind the first aortic branch, 6 mm in diameter, and post-stenotic dilatation was registered (Figure 1A). Across the pigtail catheter, we placed an Amplatzer stiff wire and an 11F long sheet. The stenosis was resolved using a 22mm long uncovered Cheatham Platinum (CP) stent crimped on a 12 mm Balloon-in-Balloon (BiB) catheter with additional flaring of the distal stent part (Figure 1B and C). The stent was manually crimped on a vacuumised balloon. The stent was inflated manually at 5 atmospheres. During the stent implantation, an overdrive external pacemaker (wire was placed into the right ventricle anterograde across the right femoral vein) was used (Video 1). Control angiography relieved satisfying anterograde flow across the aorta and aortic branches (Figure 1D). The peak-to-peak gradient after stenting was 10 mmHg. The diameter of the transverse aortic arch after stent deployment was 11 mm. Cephazolin was given 1 h before the procedure and at eight-hour intervals (total of three doses). The patient was discharged the day after the procedure after transthoracic echocardiography with antiplatelet therapy.

Physical examination and echocardiography findings immediately and 3 months after the intervention revealed normal pulses, arterial pressure, and aortic blood flow.

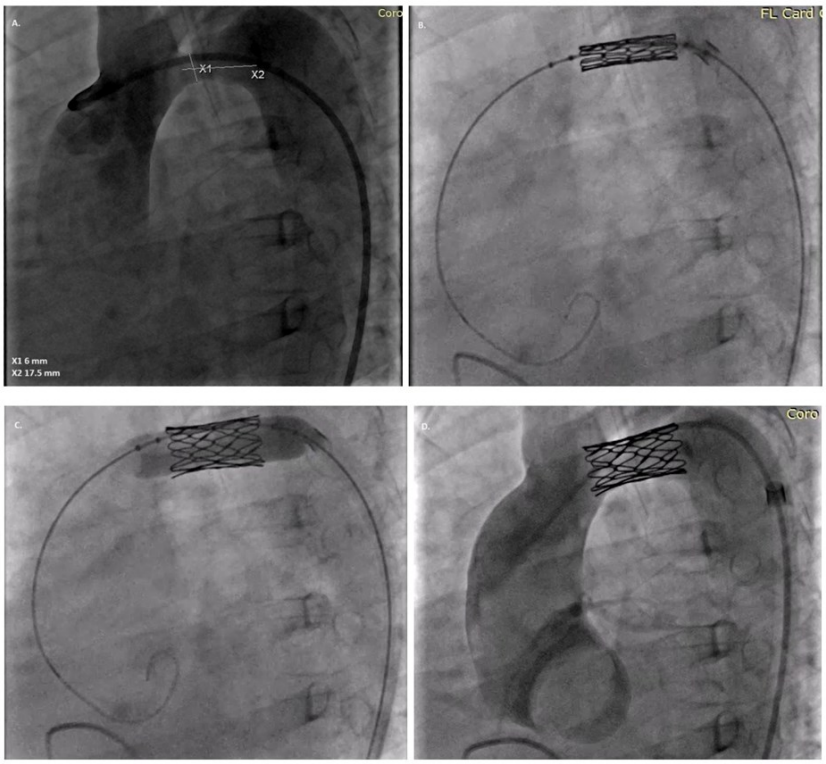
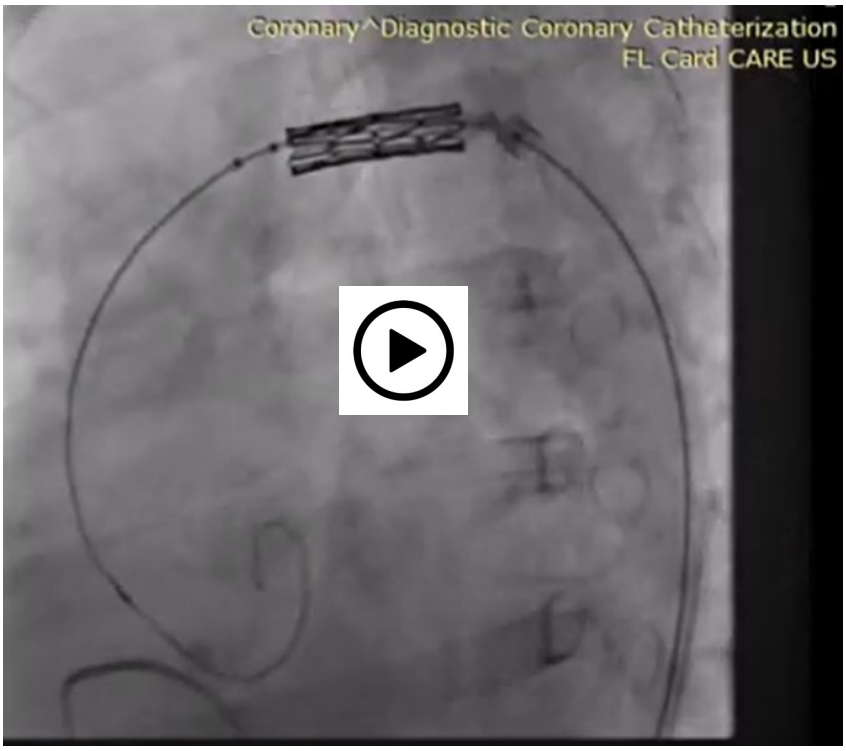


Figure 1: *A. Aortography relieved tubular hypoplasia of aortic arch, with localized narrowing right behind the first aortic branch; B. Two-stage stent implantation process using inner balloon of Balloon-in-Balloon catheter (BIB); C. Two-stage stent implantation process using outer balloon of BIB catheter; D. Control aortography after the stent implantation.*



Video 1. *Two-stage 22mm long uncovered Cheatham Platinum (CP) stent implantation process using outer balloon of BIB catheter. During the stent implantation, an overdrive external pacemaker (wire was placed into the right ventricle anterograde across the right femoral vein) was used.*

Discussion

In the last decade, percutaneous stent placement has become the standard for treating CoA after childhood. However, stent placement as a treatment for aortic arch hypoplasia has only been described in small series and case reports (5-9).

The aortic arch surgery is extensive and mutilated, which demands circulatory arrest or isolated cerebral perfusion. This is related to higher rates of neurological deficits, mortality rates and longer intensive care units (ICU) and in-hospital stays. On the other hand, patch plastic increased the risk of aneurysms and directions in the follow-up period (10).

We deployed a CP-uncovered stent in the hypoplastic tubular aortic arch and discrete stenosis in the patient with DiGeorge syndrome who underwent an IAA operation in the neonatal period. Some authors presented transverse aortic arch stenting in children and young adults (from 12 – 40 patients), with variable numbers of patients previously operated because of IAA (1 – 4 patients), but there is no data about patients with DiGeorge syndrome (5-9).

Due to the possibility of a stroke and subclavian steal phenomenon, we have used the uncovered CP stent, which was placed between the two aortic arch branches. Although the narrowest part of the arch was just behind the first aortic arch branch, we estimated that the risk of aortic dissection was lower than partially covering the common origin of the truncus brachiocephalic and left carotid artery. CP stent is preferred when the ends of the stent might protrude over the origin of a neck vessel, while the distal end struts of a unique 'zig' pattern can be opened or flared separately if necessary (8). Pushparajah et al. presented 21 patients who underwent transverse arch hypoplasia stenting; one of those patients was similar to ours and had complete surgical corrected IAA with VSD. The covered stents were used in 7 patients, while all other stents with overlapping head and neck vessels were bare metal stents (5). Warmerdam et al., in their series of 12 patients, used only different kinds of bare metal stents (6). In the study by Boshoff et al., the highest number (15/21 patients) of CP stents were deployed (8). In this study, one patient operated due to IAA (8). Holzer et al. used covered CP stents only in two patients (9). On the other hand, Pushparajah et al. used covered stents in complex aortic CoA when a left subclavian artery (LSA) was part of the CoA. Consequently, the origin of LSA was completely covered in three patients and partially in five patients without complications (5). Boshoff et al. electively completely crossed the origin of the left subclavian artery with the stent in four patients without any clinical complications in short-term follow-up (8). Although we wanted to avoid overlapping with the aortic arch branches, we decided on a 22 mm long CP stent, while the length of the hypoplastic region was estimated at 17 mm. The longer stent could protrude in the dilated descending aorta, rearrange arch architecture, and overlap the left subclavian artery.

We used overdriving pacing to achieve the correct position of the stent. A key aspect of successful TEVAR is transient hypotension during stent deployment at the proximal landing zone. Rapid and reversible reduction of systolic blood pressure to less than approximately 50-60 mm Hg helps the interventional cardiologist accurately deploy the stent and achieve optimal outcomes for the patient (11).

The stent was inflated manually, allowing gentle and gradual expansion to exceed 5 atm. This left us full control in deciding to dilate a pressure-resistant waist. Stent overdistension of such pressure-resistant hypoplastic segments could severely damage the vessel wall, with a higher risk for transmural aortic tear or rupture (8).

After the stent deployment, the peak-to-peak gradient decreased from 32 to 10 mmHg. The authors described the significant reduction in peak-to-peak gradient immediately after the procedure (5-9).

Our patient did not have any complications after stent deployment. By choosing an adequate type and length of the stent, we did not cover any of the aortic arch branches. On the other hand, by using an external pacemaker, we avoided stent migration. Pushparajah et al. described three major complications, including stent migration in two patients and a stroke in one of these two patients. Stent migration further highlights the need for rapid right ventricular pacing during stent deployment in lesions. Some other authors described only minor complications (5,7,8). 30.9% of periprocedural events were described in the study by Holzer et al., but they included critically ill end ECMO patients (9).

Conclusion

Although technically challenging, carefully analyzing the arch and vessel morphology allows the dilation of the transverse aortic arch hypoplasia and stenosis using a bare metal stent with good angiographic and hemodynamic results and without complications.

Conflict of Interest

The authors have indicated they have no potential conflicts of interest to disclose.

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