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Difficult to treat recurrent stenosis of the aorta

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Abstract

The risk associated with repeated treatment of aortic stenosis is as high as 5% and increases to as much as 25% in complex heart diseases. Among the methods that are commonly accepted and used in the treatment of recurrent aortic stenosis are balloon dilatation and stent implantation. In this study we describe five patients with recurrent stenosis of the aorta treated with stent implantation. The short-term results of such treatment are promising. However, in some cases it is only palliative in character and does not completely resolve the problems arising from congenital heart disease. (Cardiol J 2007; 14: 186–192)

Key words: recurrent stenosis of the aorta, stent implantation, children

Introduction

The risk associated with repeated treatment of aortic stenosis is as high as 5%. It increases to as much as 25% in complex heart diseases [1]. Restenosis of the aorta diagnosed after the first operation may appear in the previously treated area, at the end of the patch or at a different point from the one originally operated on. The last case mentioned is often observable when coarctation of the aorta is accompanied by a hypoplastic transverse aorta [2].

Among the methods that are commonly accepted and used in the post-surgical treatment of recurrent aortic stenosis are balloon dilatation and stent implantation [3, 4]. The most common early complications described in the literature, such as bleeding, extraperitoneal haematoma, stent dislocation, balloon rupture or dissection of the aorta by an aneurysm, can be successfully treated [4–6].

The aim of the study was to present and analyse five case reports of patients with recurrent aortic stenosis treated with stent implantation. Additionally, the report focuses on the possible difficulties and complications of the above-mentioned method.

Description of five cases of the treatment of recurrent aortic stenosis in children

The general characteristics of the patients are presented in Table 1.

Case 1

A 17-year-old boy, weighing 63 kg, who had undergone homograft implantation into the aorta, constituted a therapeutic problem because of arterial hypertension of up to 210/60 mm Hg, despite the administration of four anti-hypertensive drugs.

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Table 1. General characteristics of the patients.

No.	Sex	Diagnosis	Operation	Age of operation	Age of balloon dilatation	Weight	Age of stent implantation	Weight	Observation time
1	M	Supravalvular Ao stenosis, homograft stenosis situated distally	Homograft	10 years	0	0	17 years	63 kg	3 years
2	F	Taussig-Bing, CoA (aneurysm aortic bulb)	End-to-end	1 week	3 years	16 kg	8 years	32 kg	6 months
3	M	Williams, dist. reCoA, aneurysm	Dacron patch	6 years	0	0	17 years	35 kg	6 months
4	M	Bicuspid Ao valve, CoA, MI II	End-to-end	3 months	0	0	14 years	69 kg	6 months
5	M	Bicuspid Ao valve, CoA	End-to-end	4 months	0	0	7 years 9 years	26 kg 32 kg Dilatation	2 years 6 months

M — male, F — female, Ao — aortic, CoA — coarctation of the aorta, dist. reCoA — recoarctation situated distally, MI — mitral insufficiency

At the age of 10 he had undergone surgery for aortic stenosis, with the homograft implanted from the annulus of the aortic valve above the orifice of the left subclavian artery (LSA). Immediately after the operation the therapeutic results were satisfactory. However, shortly afterwards a relapse into arterial hypertension was observed. Echocardiographic examination showed aortic insufficiency grade III and a pressure gradient up to 96 mm Hg in the descending aorta. Angiography revealed aortic stenosis of 6.8 mm in diameter, situated distally to the subclavian artery orifice (the arteries deviating from the aortic arch were sewn onto the homograft). The anastomosis of the homograft and the hypoplastic descending aorta close below the LSA was 12 mm in diameter. The stenosis was successfully treated with the implantation of an 8 ZIG 34 mm stent. Arterial hypertension was reduced and aortic insufficiency decreased to grade II.

The patient was described in a previous report [7]. Three years after stent implantation aortic insufficiency has been graded as III and the pressure gradient in the descending aorta estimated to be up to 80 mm Hg. The patient is still receiving treatment with four anti-hypertensive medicines and his systemic blood pressure remains between 118/33 and 160/40 mm Hg. The patient's general condition is good.

Case 2

An 8-year-old girl weighing 32 kg with recurrent stenosis after a single-stage operation for coarctation of the aorta and Taussig-Bing by the arterial switch method was admitted to the Paediatric Centre in Poznań because of developing aortic stenosis.

The defect had been diagnosed in the neonatal period. At the same time a ventricular septal defect was closed and the coronary arteries were reimplanted. Owing to disproportion between the aorta and the pulmonary artery, the aortic bulb was enlarged with a pericardial patch. The aortic stenosis was dilated with the end-to-end method. Because of prolonged emphysematic changes in the left lung, bronchoscopy was performed, which revealed extrinsic pressure on the left main bronchus. When the patient was 7 months old, the aorta, which was pressing on the bronchus, was accessed via a left thoracotomy and pulled forward away from the bronchus. Over the following 3 years a murmur, increasing up to 3/6, was heard in the area of the sternum. A trans-thoracic echocardiogram (TTE) examination revealed an increase in the pressure gradient up to 56–60 mm Hg in the aorta behind the brachiocephalic trunk, an extended aortic bulb and aortic insufficiency grade I. In August, 2001 the patient qualified for cardiac catheterisation. Examination revealed stenosis of the transverse aorta up to 9.8 mm located distally to the left cervical artery, stenosis near the LSA up to 5.4 mm and a post-stenotic extension of the descending aorta up to 14 mm. The patient underwent a Tyshak 10 mm × 2 cm balloon dilatation of the aorta, which resulted in a slight decrease in the aortic pressure gradient (43–50 mm Hg). Over the following five years, however, the pressure gradient was again on the increase. TTE examination revealed slight left

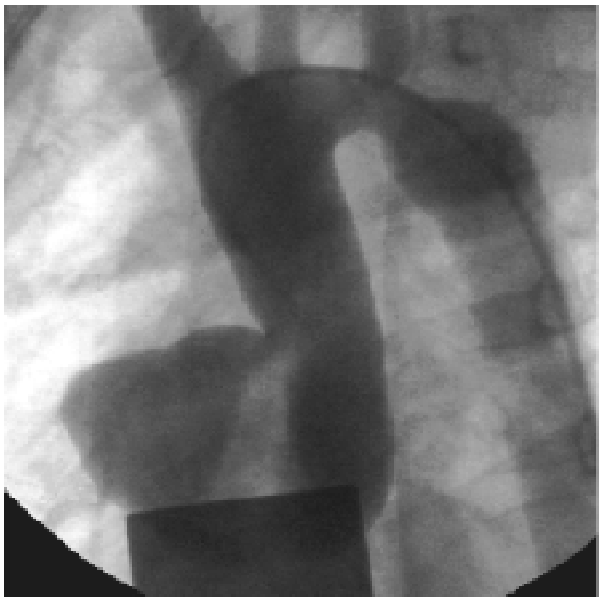

Figure 1. Stenosis of the transverse aorta. Aortic bulb dilated with a pericardial patch (the calibrated measure is 30 mm).

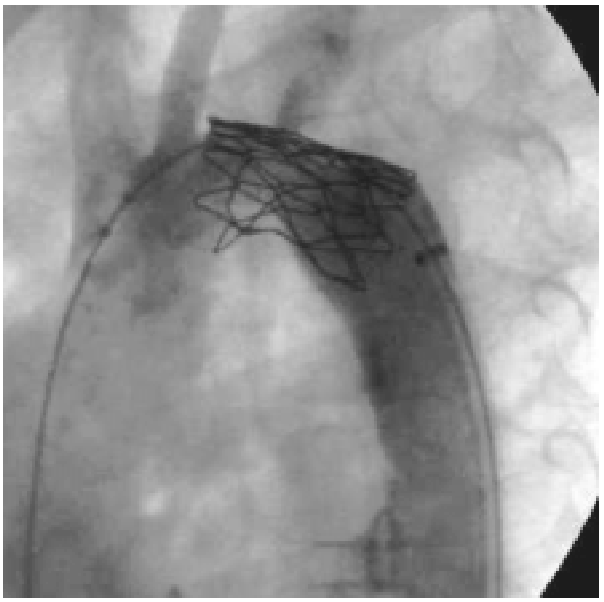

Figure 2. The transverse aorta treated with a CP-stent.

ventricular dominance, a haemodynamically insignificant residual defect, normal left ventricular contractility, aortic insufficiency grade II and a pressure gradient of 62.23 mm Hg in the aorta. ECG examination showed right bundle-branch block.

Given the results presented above, we considered further dilatation of the aortic bulb perilous. There was apprehension concerning the development of aortic insufficiency and, at the same time, awareness that surgical intervention would induce a significant risk factor. On consultation with Dr. Qureshi, we qualified the patient for another catheterisation and planned stent implantation. Aortography revealed stenosis of the transverse aorta (up to 12.4 mm) located distally to the cervical artery, another (up to 8 mm) near the LSA, post-stenotic extension of the descending aorta (up to 16 mm) and a significantly extended aortic bulb (Fig. 1). A CP Z28 stent mounted on the BIB 14 mm × 4 cm × × 8 F balloon was inserted into the aorta through a Mullins 11 F long sheath. The sheath was mounted on an Ultra Stiff 0.035 guidewire and inserted into the descending aorta below the planned stent implantation site. The stent was inserted as far as the origin of the left cervical artery and then extended (Fig. 2). During the implantation no complications were observed. After the procedure the general condition of the patient was good and ECG examination showed ST-T negative waves in leads II and III as well as in all chest leads. Echocardiography showed normal left ventricular function and dimensions. The Doppler velocity through the stent was 2.8 m/s and the gradient was 32 mm Hg. The patient was asymptomatic and there were no indicators of cardiac muscle damage (CK 129 LDH 501, troponin < 0.20) and so the patient was discharged home 10 days after the procedure without any additional tests.

Two months later the ECG reverted to normal, similar to that prior to stent implantation. A myocardial perfusion scan was normal.

Case 3

A 17-year-old boy weighing 35 kg was diagnosed with Williams syndrome and recurrent stenosis after surgery for aortic coarctation performed at the age of 6. During the procedure a hypoplastic descending aorta had been discovered.

Owing to arterial hypertension up to 200/96 mm Hg, which was unresponsive to treatment (metoprolol, captopril, nirendypine and furosemid), and a gradient in the descending aorta which reached 120 mm Hg, the patient qualified for an interventional surgical procedure. At the age of 16 he had undergone aortographic examination in Wrocław. The examination revealed stenosis of the transverse aorta up to 10 mm in the area where the proximal part of the Dacron patch had been implanted. The diameter of the transverse part of the aorta behind the brachiocephalic trunk was 13.4 mm. The aorta, which was 33 mm long, was dilated up to 17.3 mm at the place where the patch had been implanted. At the distal part of the patch there was a stenosis

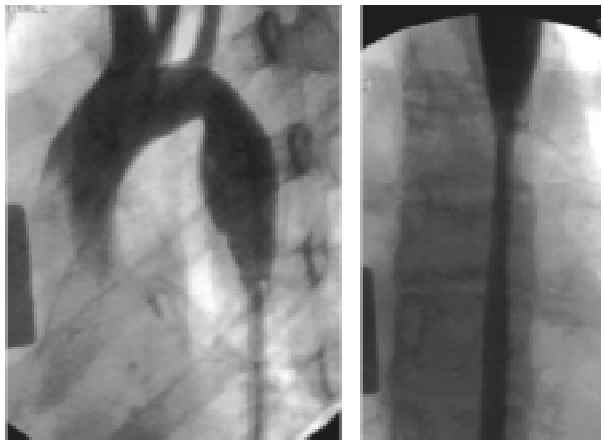

Figure 3. Stenosis of the transverse aorta in the proximal segment of the Dacron patch; post-stenotic dilatation of the aorta in the patch area. Significant stenosis at the edge of the patch.

reaching 5 mm. The aorta at the level of the diaphragm was 7.5 mm (Fig. 3). Because of the length of the hypoplastic aorta, as well as the lack of accurately sized stents, a diagnostic procedure alone was performed. On consultation with Dr. Qureshi, the boy qualified for an interventional surgical procedure. Aortography revealed significant stenosis of the descending aorta at the end of the patch (aneurysmatic aortic dilation) up to 4–5 mm and a gradient of 30 mm Hg. General anaesthesia and mechanical ventilation caused a decrease in the gradient as measured by echocardiography. There was yet another stenotic place at the proximal end of the patch, but no pressure gradient was found there. In order to check the significance of the stenosis and the reaction to dilatation a 12 mm × 3 cm Tyshak balloon was introduced further into the aorta. The vessel was dilated with a controlled pressure of 1–1.5 atm. It dilated fully without any residual incision on the balloon, yet on deflation the stenosis recurred. Next, a 3910P Genesis stent mounted on a 12 mm × 4 cm × 7 F PowerFlex balloon (Fig. 4) was implanted. The procedure brought about very satisfactory therapeutic results. The pressure gradient dropped to 7 mm Hg.

In the follow-up period the boy has been feeling well. Arterial pressure has been recorded up to 140/80 mm Hg. Hypotension medication (metoprolol, furosemid) has been administered. TTE examination has revealed a gradient of 35–40 mm Hg.

Case 4

A 14-year-old boy weighing 69 kg and diagnosed with a bicuspid aortic valve and recurrent

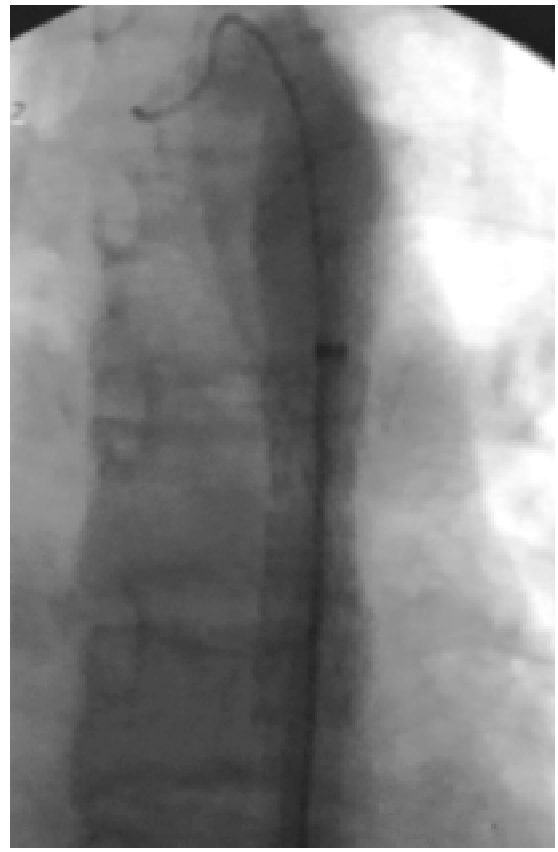

Figure 4. The aorta dilated with a stent (Case 3).

aortic stenosis, who had been operated on at the age of 3 months, was admitted to the Paediatric Centre because of arterial hypertension of 180/85 mm Hg, despite the administration of anti-hypertensive medicines (metoprolol, amlodypine). TEE examination revealed a gradient in the descending aorta reaching 90 mm Hg and aortic insufficiency grade II. The boy was qualified for invasive diagnostics. Aortography showed aortic stenosis up to 9.2 mm behind the LSA at a length of 33.6 mm. The diameter of the aorta behind the carotid artery was 14.8 mm and the descending aorta was dilated to 16.3 mm. In co-operation with Dr. Qureshi, we introduced an 8Z28 CP stent through a long 12 F Mullins sheath and expanded it using a 16 × 4.0 cm × 9 F BIB balloon. During the inflation of the balloon we used ventricular stimulation of the right ventricle in order to decrease the pressure in the aorta. The stent was not fully expanded and therefore we exchanged the balloon for a high pressure 14 mm × × 3.0 cm × 10 F Mullins. We encountered serious difficulty in retrieving the balloon and returning it to the sheath, but the procedure was successful.

Six months after the stent implantation the patient's blood pressure dropped to 140–150/60 mm Hg,

accompanied by the same hypotension treatment. In TEE the gradient has been about 73 mm Hg. The blood flow through the abdominal aorta has an arterial pattern. We cannot exclude stenosis at the distal part of the stent. An magnetic resonance imaging examination is difficult to perform because of the patient's slight mental retardation and claustrophobia.

Case 5

A 9-year-old boy weighing 32 kg, who had been diagnosed with bicuspid aortic valve and recurrent stenosis and had undergone an end-to-end operation at the age of 4 months and stent implantation at the age of 7, was admitted to the Paediatric Centre for further extension of the stent.

At the age of 4 he had been catheterised because of hypertension reaching 140/70 mm Hg and a gradient shown by TEE examination to reach 80 mm Hg. Aortography revealed stenosis up to 5 mm behind the LSA at the point previously operated on. The aorta at LSA level was 10 mm, whereas distally, behind the stenotic site, it was 9.1 mm, owing to post-stenotic dilatation. A balloon dilatation was performed using the 8 mm × 2.0 cm Tyshak balloon. At first the therapeutic results seemed satisfactory, but soon after the procedure a recurrence of hypertension was observed and the gradient in TEE was 58 mm Hg. After another catheterisation performed 3 years later significant stenosis was diagnosed, prompting the decision to implant a stent. Aortography revealed stenosis up to 6mm. at the same point behind the LSA. The diameter of the aorta measured behind the LSA was 11.5 mm and there was distal post-stenotic dilatation to 12 mm. A CP ZIG 39 stent mounted on a 14.0 × × 4.5 cm BIB balloon was implanted and a 12 mm × × 4.0 cm PowerFlex balloon was then used to extend the middle and the top parts of the stent once again. As a result, the aorta was dilated up to 8 mm and the gradient in TTE study was 30 mm Hg. Further redilatation of the stent was planned to correspond to the patient's growth.

Over the following two years the gradient in the stent increased to 60 mm Hg, despite the patient being administered enalapril. Another aortography revealed an 8 mm stenosis in the central part of the stent. In distal parts the diameter of the stent and the aorta was 13 mm. In co-operation with Prof. Weil we re-extended the critical part of the stent up to 10.8 mm using a 12 m × 4 cm PowerFlex balloon. Haemodynamic examination revealed that the gradient measured through the stent was 11 mm Hg, whereas in a TTE study performed after the procedure it was 29 mm Hg. The clinical condition of the child is good and he has blood pressure of 100/60 mm Hg.

Discussion

The treatment of aortic recoarctation by stent implantation is a commonly accepted method of treatment [3, 6, 8]. When stent implantation is planned, allowances have to be made for the possibility of treating complications as well as the necessity of implanting a covered stent [4].

In the case of infants and young children, balloon angioplasty is advised as the first stage of treatment of aortic recoarctation [4]. However, these procedures are not always successful [9]. In VACA (Valvuloplasty and Angioplasty of Congenital Anomalies registry) studies the results of balloon angioplasties of aortic recoarctation are better when the interval between the two procedures is shorter [10].

We applied the same strategy to two of our patients (Cases 2 and 5). Balloon angioplasty of the coarcted aorta was performed at the ages of 3 and 6, whereas the stents were implanted in at the ages of 8 and 7 respectively, when the stenosis of the aorta had increased. Stent dilatation is possible in the follow-up period and has to correspond to the patient's growth [4]. We performed such a dilatation in one child using a balloon designed for high pressure (higher than used originally for stent implantation).

In the literature there are also reports of stent implantation in the transverse aortic arch [11, 12]. Surgical dilatation of the transverse arch is more difficult than the analogous treatment of the descending aorta. The operation is performed under hypothermia, using sternothomy [13]. Our second patient, diagnosed with coarctation of the transverse aortic arch, had already undergone two operations. Implanting a stent in the transverse aortic arch allowed her to avoid yet another operation. A small residual narrowing of the stent may be redilated in the follow-up period, corresponding to the child's growth. Owing to the fact that the stenosis contained the origin of the LSA, the stent length had to be correctly adjusted so that it would dilate the most important segment of the narrowing, that from the origin of the left carotid artery to the post-stenotic dilatation of the descending aorta. We did not hesitate to cover the opening of the LSA with a stent; aortography revealed a normal inflow of blood into this vessel.

There are many studies reporting the dilatation of the aortic bulb after an "arterial switch"

operation. The reason for such dilatation remains obscure; so far it has not been clearly defined [14]. There is aortic valve insufficiency, ranging from moderate to significant, in 3% of these patients. However, it is seems that the disturbed outflow from the aorta may increase aortic insufficiency as well as aortic bulb dilatation at the pericardial patch.

Studies do not report on the ST-T segment changes after a complication-free stent implantation in the aorta, yet none of the patients described had been treated with the "arterial switch" method. Temporary ST-T segment changes observed during interventional procedures are assumed to be caused by air accessing the coronary arteries [15]. Perloff describes abnormal flow through dilated extramural coronary arteries with a changed coronary microcirculation response to the vascular endothelial growth factor and nitric oxide in cyanotic congenital heart diseases [16]. Other authors emphasise abnormal pulsatile flow through the aorta in syndromes with disturbed elastin function (Marfan Syndrome) [17]. In our patient (case 2) the temporary ST-T segment changes might have been related to decreased flexibility and compliance of the aortic bulb after its augmentation with a pericardial patch caused by pressure changes after stent implantation, when there may be a decrease in the pressure gradient between the ascending and the descending aorta. After two months the ST-T segment condition returned to normal. Normal flow in the coronary arteries was confirmed by radioisotope study.

Numerous authors report the creation of aneurismal aortic dilatation at the patch concomitant with narrowing of the transverse aortic arch [18, 19]. In our third patient we found aneurismal patch dilatation. There was only a moderate narrowing of the transverse aortic arch, but a significant narrowing was found at the distal end of the patch in the descending aorta. This narrowing seriously hindered the blood outflow from the segment of the aorta dilated with the patch. Like the doctors from other medical centres who had previously examined the child, we were afraid that the complete dilatation of the aorta would require implantation of a number of stents. However, a measuring test using a low pressure balloon enabled us to define precisely the segment of most significant stenosis and to evaluate the compliance of the aortic wall. In this particular case we decided to use a Genesis stent, which is stiffer than a CP stent.

In complex congenital heart diseases, particularly in those accompanied by a hypoplastic aorta, stent implantation is not equivalent to restoring normal conditions, as was the case in the first and the fourth patients presented above. However, a significant decrease in arterial blood pressure makes the patient's life more comfortable and facilitates pharmacological treatment. In four of our patients there was a significant drop in arterial blood pressure. High blood pressure in the ascending aorta affects the volume of the regurgitation flow through the aortic valve. The aortic valve insufficiency grade decreased in two patients.

There are reports of the attractive short-term effects of stent implantation in the aorta [20]. At the same time the necessity of further examination, including clinical examination, echocardiography and multislice computed tomography or magnetic resonance imaging, is also emphasised [21, 22]. These examinations have been scheduled for the patients presented in this report.

We managed to implant stents safely in our patients, in all of whom it was evident that a high risk factor would accompany any potential surgical manoeuvres. This enabled the detrimental symptoms of recurrent aortic stenosis to be reduced and further surgery thus avoided.

It has to be pointed out that the satisfactory short-term results of the percutaneous interventional aortic stenting were to a large degree obtained thanks to the experience and skills of the interventional cardiologists who assisted us during the procedures.

Conclusions

1. The short-term results of stent use in the treatment of post-surgical aortic recoarctation are satisfactory.
2. In some cases interventional treatment using stents has a palliative character and does not completely resolve the problems arising from congenital heart disease.

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