Aortic Coarctation Treated by PTA and Stenting: a Case Presentation and Literature Review

Angela GEORGESCU; Eustaquio ONORATO; Silvia NICOLAE; Serban BALANESCU

a Department of Cardiology, Monza Hospital, Bucharest, Romania
b Cardiovascular Department, Humanitas Gavazzeni Hospital, Bergamo, Italy
c Department of Radiology, Monza Hospital, Bucharest, Romania
d Department of Angiography and Endovascular Interventions, Monza Hospital, Bucharest, Romania

ABSTRACT

A 13 year-old boy was diagnosed with simple aortic coarctation and a bicuspid aortic valve. Ascending aorta was mildly dilated while aortic arch was smaller, but not hypoplastic. A decision to treat him was made depending on the clinical finding of systemic hypertension in his young age and a baseline trans-stenotic echographic gradient of 48 mmHg. Direct stenting with a covered Cheetham-Platinum balloon-expandable stent was performed with no complications. Despite immediate good angiographic and hemodynamic result with marked decrease of invasively measured gradient, consecutive echographic follow-up demonstrated progressive increase of the gradient in the first 9 months after the index procedure. This resulted in repeated aortic PTA with a larger balloon inside the previously implanted stent-graft. Optimal angiographic result was obtained, the patient remained symptom-free and no antihypertensive therapy was necessary thereafter.

Modern treatment of simple aortic coarctation can be done in the cath-lab in appropriately selected cases. Stent implantation should be considered in all cases to avoid dissection and early elastic recoil. Long term follow-up may evidentiate the need for repeated PTA procedures in some cases.

INTRODUCTION

Aortic coarctation is a discrete stenosis of the juxta-ductal thoracic aorta distal to the left subclavian artery. It has a prevalence of 1 out of 3000-4000 live births and may be associated with other congenital heart disease like bicuspid aortic valve, congenital aortic stenosis, persistent ductus arteriosus or ventricular septal defect. It represents about 5-8% of all congenital heart disease (1).

Neonates diagnosed early with aortic coarctation have more frequently complex anatomy and more associated malformations. Girls with aortic coarctation should be screened for Turner syndrome due to frequent association between the two.
Coarctation patients with secondary hypertension are at a lifelong risk of aortic dilation, rupture or dissection even long after successful repair. In these cases proper serial follow-up should be done although no clear recommendations exist.

CASE REPORT

We present the case of a 13 year-old boy referred to our Hospital by the school doctor who discovered an elevated blood pressure at rest (150/90 mmHg) on routine examination. The patient was asymptomatic at rest and described sometimes headache on exertion.

His mother went through cardiovascular surgery for aortic coarctation at the age of 21 and thereafter she had two normal pregnancies: first a healthy girl and subsequently our male patient who was three years younger.

On clinical examination the patient showed normal physical development (168 cm high and 63 kg of body weight), with a clear chest auscultation and normal breathe sounds. A heart rate of 59 beats per min and a grade II/VI mid-systolic murmur located at the apex and irradiated to the left interscapulovertebral area were noted at cardiovascular examination. The rest blood pressure was 145/100 mmHg in the left arm and 140/90 mmHg in the right arm. A weak pulse was felt in both femoral arteries. All other details of clinical examination were interpreted as normal.

A standard 12 leads EKG was normal at rest, in sinus rhythm and no signs of left ventricular hypertrophy or strain.

Transthoracic echocardiography revealed a bicuspid aortic valve with no significant systolic gradient, grade I aortic regurgitation and a nondilated LV with slight concentric hypertrophy and normal systolic function. Suprasternal view used to visualize aortic arch and the isthmus confirmed coarctation of the aorta with a maximum trans-stenotic gradient of 44 mmHg and a diastolic gradient (Figure 1A). For more accurate characterization of aortic anatomy and precise measurement of minimal lumen diameter an aortic angio-CT scan was performed. It confirmed the coarctation of the descending thoracic aorta located 2 cm below the origin of the left subclavian artery with a 6 mm minimal lumen diameter; this was interpreted as a reduction of 84% compared to distal thoracic descending aorta (Figure 1B). The length of aortic narrowing was rather short, of 5 mm length. No post-stenotic dilatation of the descending aorta was noted. There was a large collateral circulation path at the postero-superior chest wall.

FIGURE 1. Transthoracic echocardiography showed a 44 mmHg trans-stenotic gradient in the descending aorta (Figure 1A). Angio – CT examination of the aorta precisely located the obstruction (white arrow) and demonstrated an appropriate distance from the ostium of the left subclavian artery for safe stent placement (Figure 1B). Aortic arch was half the diameter of the ascending aorta (yellow arrow).
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THERAPEUTIC OPTIONS

A recent American Heart Association scientific statement defined the indication for stent placement in native and recurrent aortic coarctation (2). However, the decision as to which method should be used for the treatment of coarctation out of the options of surgical, balloon angioplasty and stent placement remains controversial. In the experience of the CCISC the vast majority would favour either balloon angioplasty or surgical treatment of the coarctation segment in patients weighing less than 25 kg for late-onset coarctation (i.e., older than 4 years of age) (3). Once the patient grows older and weighs over 25 kg, opinions are equally divided between balloon angioplasty and stenting of the coarctation segment, with primary surgical treatment becoming less frequent (4).

A Heart Team decision was made (after thorough discussion between the cardio-vascular surgeon, interventional cardiologist and both parents) to perform interventional treatment due to its less invasive approach in a patient almost of adult size (with a class IIa indication in the American Guidelines). Aortic PTA with stent implantation was done in the regular angiography suite without any complication.

INTERVENTIONAL PROCEDURE

A sentinel pig-tail 6F catheter was inserted by left radial approach in the aortic arch immediately upstream from the coarctation, under mild sedation with midazolam. Right common femoral approach was obtained with an 8F introducer sheath. Diagnostic angiography in digital subtraction was performed and trans-stenotic maximum instantaneous gradient was measured at 50 mmHg.

A Back-up Meier SuperStiff 0.035” 260 cm long guide-wire was then deployed in the ascending aorta. A 10F Mullins sheath was advanced over the stiff guide wire and left in the aortic arch. A covered Cheetham-Platinum NuMed balloon-expandable stent was mounted on a 14x40 mm dedicated Tyshak BiB balloon (balloon-in-balloon) and was advanced in the Mullins sheath in the aortic arch. The sheath was then carefully retracted exposing the stent at the maximum level of the stenosis taking care to avoid the ostium of left subclavian artery. Contrast injections through the sentinel pigtail catheter were used to precisely position the stent (Figure 2A). The covered stent was then expanded at the nominal pressure aiming to double the baseline minimal luminal diameter of the coarctation (12 mm vs 6 mm at baseline), with correct stent expansion and good angiographic final result, 10% residual stenosis and a pressure gradient less than 5 mmHg.

In the first day after the procedure blood pressure increased up to 190 mmHg; a paradoxical hypertensive reaction is frequently described after correction of aortic coarctation, mainly after surgical intervention. After discharge blood pressure remained normal without medication for 2 months. Slightly increased BP was noticed thereafter up to 145/90 mmHg and the patient resumed treatment with 2.5 mg bisoprolol od.

A control angio-CT after intervention showed the stent correctly positioned. Despite low final pressure gradient across the stented segment, post-procedural ultrasound demonstrated a persistent maximum gradient of 32 mm in the descending thoracic aorta. In the first two months after the dilatation, the echo gradient in the descending aorta remained stable; 3 months after the procedure it has risen to 46 mmHg, but without diastolic gradient.

Nine months after aortic PTA and a height increase of 10 cm and 6 kg in body weight we decided to restudy the patient in the cath-lab and measure aortic gradient. Aortic angiography was performed by right femoral approach documenting the correct position of the previously implanted stent, without in-stent restenosis or recoil. The pullback “peak to peak” pressure gradient between the aortic arch and descending aorta was 31 mmHg (116/77 mmHg in the ascending aorta and 85/69 mmHg in the descending aorta. We decided to dilate the CP stent-graft previously implanted to reduce the gradient; considering that stent implantation was done with a Tyshak 14mm balloon, which meant doubling the initial aortic diameter, we chose a 18x40mm NuMed balloon at the nominal pressure of 8 atm x 4 (Figure 2B). The residual pullback “peak to peak” gradient was 9 mmHg. Should the use of an 18x40 mm balloon be insufficient to significantly lower the peak-to-peak gradient, a higher diameter balloon would have been employed and the stent repeatedly dilated until a non-significant gradient would have been obtained. Echocardiography after balloon dilatation showed a maxi-
mum velocity of 2.98 m/sec, with a maximum gradient of 35 mmHg.

At 3 months after stent re-expansion the patient is asymptomatic, with normal blood pressure without antihypertensive treatment, while the same gradient was recorded by echocardiography at the stent level.

The echocardiographic persistence of the pressure gradient in the descending aorta raise the question of its origin: insufficient stent expansion with residual stenosis (excluded since registration angiographic gradient after the first procedure was 5 mmHg, aortic arch hypoplasia with flow acceleration generated before the isthmus and the coarctation site (the angio-CT examination revealed reduced dimensions of aortic arch, between the ostium of left common carotid and subclavian artery); or physiological acceleration intrastent. Probably a combination between the last two could be held responsible for the flow acceleration recorded by Doppler study. However, Doppler gradients are not useful for quantification, neither in native nor in post-operative coarctation. A diastolic ‘run-off’ phenomenon is presumably the most reliable sign of significant coarctation or recoarctation. In the presence of extensive collaterals the gradients are not reliable (5,6). Echocardiography should thus be used as adjunctive to clinical follow-up and should not be used to diagnose significant progression of the disease (7).

We consider this case as peculiar due to:

a) Familial association of aortic coarctation with more severe phenotypic expression in the child who also has a bicuspid aortic valve. There are case reports of families with several members suffering from aortic coarctation (8). In a systematic study of this congenital disease, familial aggregation was considered as a result of multifactorial inheritance and recurrence risks in siblings was evaluated at about 0.5% for coarctation and 1.0% for any form of congenital heart defect (9). Nevertheless, in some of the described families, aortic coarctation seems to be inherited as an autosomal dominant mutation with incomplete penetration and variable expressivity (10). As far as there is no guideline recommendation for genetic testing in aortic coarctation and literature mentions just some families of patients with coarctation, with no particular gene involved, we did not proceed with blind genetic probing.

b) The need for early balloon reexpansion, which was due either to elastic recoil of the stent-graft (which should not be significant) or to the patient grow-up (significant in 9 months). Initial stent implantation could not be done at more than the double of mini-
mal aortic lumen diameter (6 vs 12-14 mm), that can be considered as insufficient with respect to the patient size; thus post-dilatation with an 18 mm balloon 9 months after the index procedure further increased stent-graft diameter, more appropriate to the patient body size.

**DISCUSSION**

Transcatheter approach to aortic coarctation was first performed using balloon angioplasty in the 1980s (11), with intravascular stent treatment gaining wider acceptance in the 1990s (12). At many institutions transcatheter approach has become the treatment of choice for children and adults with native coarctation of the aorta (13). A recently published Cochrane review demonstrated that no randomized trials were available to compare surgery and stent repair (14). Considering the available data we have to be aware each technique had its own evolution and learning curve and there is a large difference with regard to the length of follow-up available after each treatment option, with approximately 10 years after stent repair, 20 years after balloon angioplasty and up to 50 years after surgery. Unfortunately accurate follow-up for all 3 types of treatment is limited, making it difficult to draw any meaningful conclusions as to which treatment option is superior. The issue is further complicated by uncertainty as to what constitutes an acceptable upper to lower extremity systolic blood pressure gradient after repair. Furthermore, aortic arch imaging with CT, MRI or cardiac catheterization is not routinely performed after percutaneous repair, which results in suboptimal screening for long-term complications (15). Due to these limitations decision-making remains challenging, and the choice for the most effective and safe treatment option may depend on patient’s age and individual clinical scenario. In a study of CCISC stent patients had significantly lower acute complications compared with surgery patients or BA patients, although they were more likely to require a planned reintervention with no differences observed among the 3 groups regarding the need for performing unplanned reinterventions. At short-term and intermediate follow-up, stent and surgical patients achieved superior hemodynamic and integrated aortic arch imaging outcomes compared with BA patients (3). Surgery is considered the treatment of choice in infants and results in a mortality rate of less than 1%. Balloon angioplasty is not recommended in neonates and children due a higher re-intervention rate due to restenosis and an increased risk for aortic rupture and aortic aneurysm formation as compared with surgical repair. Aortic surgery is still the treatment of choice in children beyond the neonatal period. The main reason for which surgery favours stent repair is the increased risk of complications after stent implantation in small children that did not grow-up, usually below the age of 8-10 years. Most feared complications after surgery are paraplegia that might occur early after intervention and late development of aortic aneurysms at the treated site (1). Frequent complications after stent repair involve difficulties with sheath delivery, vascular complications, restenosis and aortic aneurysm formation and failure to adapt to the growing child and thus redilatation becomes necessary. In young children we should investigate alternative options, including not using stents in discrete coarctations and exploring further research with biodegradable and growth stents (16).

Although mortality rates are higher after surgical repair, reintervention risk remains higher after stent repair. Most reinterventions involve redilatation due to under-dilatation during the initial treatment, whereas the overall risk after redilatation remains lower as compared with after repeat surgery.

Most cardiologists use stents in adolescents and adults. The stent implantation procedure is classically considered successful if the residual gradient is less than 10 mmHg, with an improvement in diameter to >90% of the normal adjacent aortic arch vessel (13,17-19). The success rate is >90% (13,19-21), generally with no residual gradient, whereas balloon dilatation alone usually results in persistent gradient. The re-coarctation may result from remodelling of the coarctation segment and development of neointimal proliferation, stent fracture, stent recoil and physical growth with progressive mismatch between the stented segment and growing aortic diameter, if the procedure was done prior to puberty (20). Re-expansion of the stents used to treat residual or recoarctation and growth-related narrowing seems feasible, relatively safe, and largely effective (22). In the study by Duke et al. (23), although repeat dila-
tion achieved improved stent diameter and peak pressure gradient, much of the re-expansion was necessary simply to restore the original lumen size, which was reduced between the times of implantation and restudy. Stent therapy seems to be an attractive and preferred alternative to surgical or balloon therapy for treatment of aortic coarctation in the adolescent and young adult (1). Recently self-expandable non-covered stents have been used to treat aortic coarctation in adults in some tens of cases (24,25). No direct randomized comparison between the use of the CP balloon-expandable stents and self-expandable nitinol stents is available; thus no long term outcome differences between the two stent types is currently known.

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REFERENCES