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ORIGINAL RESEARCH

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Treatment of a severe distal thoracic and abdominal coarctation with cutting balloon and stent implantation in an infant: From fetal diagnosis to adolescence

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Abstract

Background and Aims: Abdominal coarctations are rare. Surgical treatment is difficult and requires re-interventions to adjust the graft material to patient growth. We report effective treatment by interventional catheterization in an infant with the concern to allow adjustment for growth and prevention of vessel damage.

Methods and Results: After the diagnosis of abdominal coarctation at 27 weeks of gestation, an infant developed hypertension (170/70 mmHg) at 3 months of age despite medical therapy. Angio CT confirmed a 2 mm diameter, 2.3-cm-long coarctation of the descending aorta. At 4 months, a dilatation was performed using a 3 mm cutting balloon and a 5 mm Opta® balloon, Cordis®. Two noncovered Palmaz® Genesis™ XD PG1910P stents were required to keep the aortic lumen open. At 15 months, an Adventa™ V12 vascular 12 × 61 mm long covered stent was implanted to exclude an aneurysm which developed between the two stents. At 3 and 9.5 years, the stents were further dilated with a high-pressure balloon to reach 11 mm aortic diameter with no residual pressure gradient, and normal blood pressure.

Conclusions: The use of cutting balloons and stent implantation is an effective way to relieve severe obstruction in middle aortic syndrome in neonates. The technical issues encountered were the need for a low profile sheath and material to avoid femoral artery damage, and the need to use stents that can be further expanded to adult size.

KEYWORDS

abdominal coarctation, children, infant, interventional catheterization, middle aortic syndrome, stents

1 | INTRODUCTION

Middle aortic syndrome is a rare clinical condition with a segmental narrowing of the abdominal or distal descending thoracic aorta,[1–3] responsible for severe arterial hypertension proximal to the aortic stenosis. Complications are gastrointestinal tract ischemia, renal failure, aneurysm rupture, cardiac hypertrophy, and arrhythmias. The etiology can be congenital, caused by a developmental anomaly in the fusion and maturation of the paired embryonic dorsal aortas or acquired, in association with Takayasu's or temporal arteritis,

neurofibromatosis, fibromuscular dysplasia, retroperitoneal fibrosis, mucopolysaccharidosis, and the Williams syndrome.

Severe hypertension is the primary indication for intervention. Clinical benefits and complications have to be balanced against the impact of the intervention. Surgical treatment entails aorto–aortic bypass, patch aortoplasty, and bypass grafting with autologous conduits. Surgical and interventional catheterization techniques are mostly described in adolescents and adults. There are few reports of interventional catheterization techniques, some with stent implantation in neonates and toddlers.[4–6] The limits of surgical treatment in newborns are technical difficulties due to the size of the patient and the adjustment with the growth of the graft implanted. The concerns for catheterization are the necessary sheath sizes for tiny baby arteries, and the possibility to dilate the implanted stent to adult size.[7,8]

2 | CASE REPORT

Prenatal diagnosis of a low thoracic 2 cm long aortic coarctation was made at 27 weeks gestational age and confirmed at birth. The patient developed severe arterial hypertension (170/90 mmHg) at 3 months of age, with 70 mmHg pressure gradient between the upper and lower limbs.

Echography showed normal cardiac anatomy and function, narrowing of the aorta at the diaphragmatic level with 8.5 m/s Doppler velocity. An angio CT confirmed a tight aortic coarctation of 2 mm diameter and 2.3 cm length (Figure 1). Ophthalmic examination did not show Lisch nodules (a nonconstant finding in neurofibromatosis before the age of 6 years).[9,10] Genetic and inflammatory screenings were not contributive.

Due to the age (4 months) and weight (6.8 kg) of the infant and the length of the stenotic part, surgeons were not keen to intervene and catheterization was performed.

Via a 4-French (F) introducer placed in the right femoral artery using ultrasound guidance, angiography showed a 1 mm diameter 3.5 cm long coarctation of the distal part of the descending aorta just above the diaphragm (Figure 1) with 73 mmHg systolic pressure gradient. Dilatation with a 5 mm Opta® Pro PTA balloon Cordis® (Opta® balloon), at 10 atm was not successful. A 3 mm cutting balloon (Flextome™Cutting Balloon™ Monorail™ 30 × 10 mm stabilized in a 6 F Cook® Flexor® Check Flow Introducer 70 cm (Flexor®sheath) allowed successful dilation of the coarctation segment. Further dilatation was performed with the 5 mm Opta®balloon, with no waist on the balloon but complete recoil of the segments at 2 mm diameter after balloon deflation. A noncovered closed cells stent expandable up to 18 mm, Palmaz® Genesis™ XD PG1910P Cordis® stent (Palmaz PG1910P), was mounted on a 5 mm Opta®balloon placed over the wire through a 6 F St. Jude Medical introducer. The balloon with the stent was pulled back in the 6 F introducer and gently inflated to flair the balloon extremities over the stent to fix the stent on the balloon. The stent was implanted through the 6 F Flexor® sheath (Cook®) after the removal of the flexor valve with a scalpel to allow stent and balloon introduction.

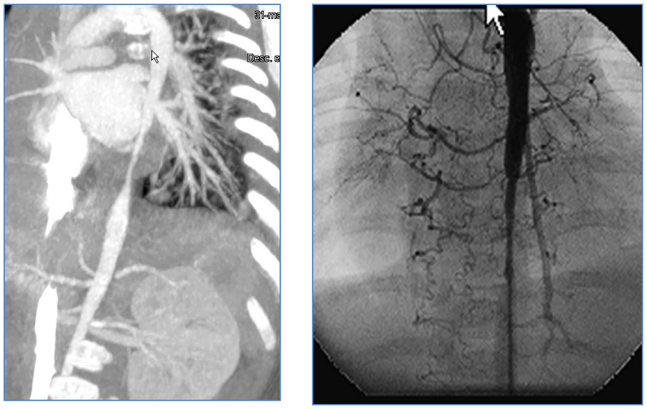

FIGURE 1 CT scan and angiography showed a tight low thoracic and abdominal coarctation with collateral vascularization.

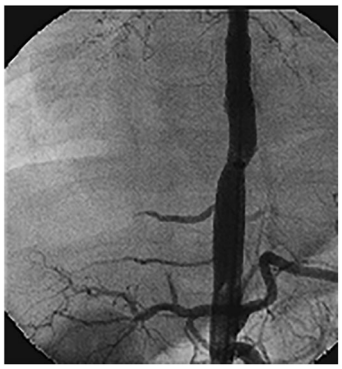

FIGURE 2 Result at the end of the first catherization procedure. A gap of 11 mm is observed in between two stents (Palmaz® Genesis™ XD [closed cells] PG1910P stents), with a 4–5 mm passage.

The 6 F St. Jude introducer was then advanced in the 6 F Cook® Flexor® Check to serve as a hemostasis valve. At full inflation, the stent migrated to the proximal part of the thoracic aorta. The stent was recovered on a 6 mm Opta® balloon and pulled back in position. The 19 mm length stent covered only the proximal part of the coarctation, and a second Palmaz® PG1910P stent mounted on a 6 mm Opta® balloon was placed in the distal part of the coarctation. Because of the migration of the first stent, there was an 11 mm gap between the two stents with a 4–5 mm aortic diameter (Figure 2) between the two stents with a 17 mmHg residual pressure gradient. The procedure was considered successful at this stage.

At clinical follow-up (FU), the patient had good femoral pulses with no blood pressure gradient between the upper and lower limbs. Medical treatment included propranolol for residual hypertension (133/65 mmHg), acetyl salicyl acid (ASA) (3 mg/kg/day) and clopidogrel (0.2 mg/kg/day). Low weight molecular heparin (LMWH) was administrated for 1 week to minimize the risk of femoral artery thrombosis and stopped after echo-Doppler showing no sign of vascular thrombosis. The patient was discharged from the hospital after 3 days, with monthly clinical and echo FU evaluations in clinics. A CT angiogram 1 month after stent implantation revealed an aortic

aneurysm between the two stents. The aneurysm diameter increased up to 12 and 15 mm, respectively, on the control CT performed 2 and 4 months after stent implantations.

A second catheter intervention was performed 11 months later (age 15 months, weight 11 kg) to implant a covered stent to exclude the aneurysm to prevent rupture.

Despite the use of low profile sheath, normal Doppler-echo control, and the use of extended heparinization, the need for large sheath diameters has led to occlusion of the right iliac artery. A 4 F left arterial access was obtained and was upgraded for a 9 F Saint-Jude introducer. There was a 9 mmHg pressure gradient, no intimal proliferation in both stents on angiography, and a 15 mm diameter–20 mm length aneurysm in between both stents (Figure 3). Both stents were dilated with a 8 mm Opta®balloon, followed by deployment of a long premounted covered stent (Adventa™ V12 vascular covered stent 12 mm × 61 mm × 120 cm) covering the aneurysm and both stents. Full expansion of the covered stent was impossible with the Adventa® balloon. A high-pressure 8 mm × 3 cm Conquest® PTA balloon (ref CQ-7583), Bard® inflated at 22 atmosphere was required to reach a 7–8 mm diameter with a 3 mmHg residual pressure gradient (Figure 4). Treatment on discharge included ASA (5 mg/kg/day), clopidogrel (0.2 mg/kg/day), and propranolol (2 mg/kg/day) for 3 months and LMWH for 1 week.

The boy was seen twice a year in a clinic with a control echo-Doppler. Further catheterization was planned based on arm–leg blood pressure difference above 15 mmHg, a diastolic extension of the Doppler gradient, a significant discrepancy between the inner lumen of the aorta in the stent and below the stent. During the third catheter intervention (age 3.2 years, weight 18 kg), and the fourth catheterization (age 9.5 years, 33 kg) the stents were further dilated with high-pressure balloons (Conquest® Bard® PTA balloon 8 and 10 mm, Atlas Gold Bard® PTA dilatation catheter 12 mm) inflated up to 28–30 atmosphere to reach full expansion and a final diameter of 11 mm with an 8 mmHg residual pressure gradient, no arterial hypertension (105/57 mmHg), no blood pressure gradient between upper and lower limbs. Medical treatment is limited to ASA (5 mg/kg/day) since the age of 21 months. The boy is now 12 years old (body weight 46.5 kg, height 150 cm). He is regularly seen on follow-up, with a perfect evolution with a normal blood pressure and no episode of visceral or renal dysfunction.

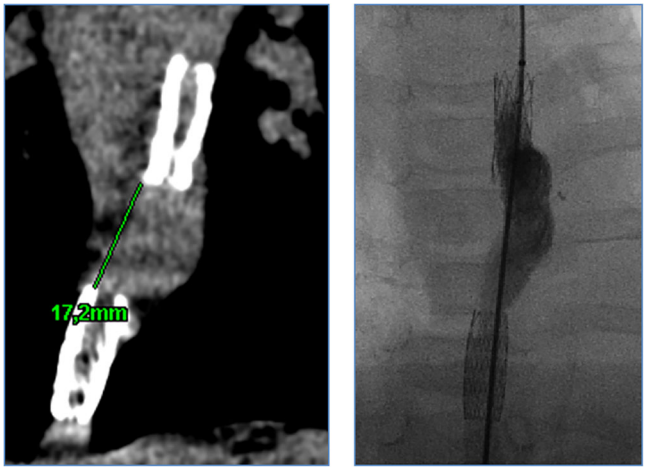

FIGURE 3 CT scan and angiography showed an aneurysm formation between the two stents in the low thoracic aorta.

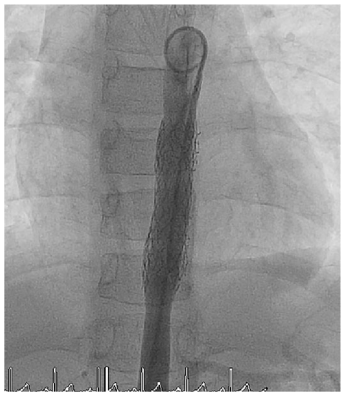

FIGURE 4 Adventa™ V12 vascular covered stent covering both stents and the aneurysm in between stents.

3 | DISCUSSION

Surgical intervention for low thoracic aneurysm is possible but difficult and considered a high-risk intervention. When intervention is performed in small infants, reinterventions are necessary in all cases to adapt the graft size with growth.[10]

Stent implantation by heart catheterization is an effective way to relieve the aortic obstruction. Limited data are available on young children, especially in babies and toddlers. Studies show an immediate reduction of systolic pressure gradient; hypertensive treatment was maintained in immediate follow-up and withdrawn later on in a few patients.[4–6]

Vessel wall integrity is supported by stenting after dilatation, minimizing the risk of aneurysm formation or dissection after balloon angioplasty alone. Major limitations for stent implantation in childhood are the diameter of the stent delivery system compared to the infant vessel,[8] the need to use stents expandable to adult size vessel, and the risk of intimal overgrowth in the stents.

The first two stents implantations were performed at 4 months of age in a 6.8 kg patient. The Palmaz® PG1910P stents, mounted on a 5 mm Opta® balloon, enters in a 6 F Flexor® sheath, an acceptable introducer size for this baby's vessel sizes given the complicated pathology. Because of the tight stenosis and despite pre-dilatation with a cutting balloon, the Opta® balloon with the stent was pushed proximally. Stabilization of the stent during the expansion of the balloon is challenging. Femoral thrombosis of the right iliac artery was observed despite LMWH treatment.

To exclude the aneurysm between the two stents and to limit the risk of intimal proliferation,[11] a covered stent was implanted at 15 months of age. The risk of aneurysm rupture was balanced upon the constraint of vascular access. For this reason, the second procedure was planned when the boy's weight was above 10 kg allowing the use of a 9 F sheath needed for the introduction of an Adventa® V12 premounted stent on an 8 mm balloon.

Preventing vessel stenosis in very young patients is a major challenge. The use of ultrasound-guided puncture is mandatory, allowing also to measure the vessel size to check dimension compatibility with the introducer. The use of deep sedation, appropriate positioning, adequate heparinization, and of stepwise predilatation before introducing a larger sheath are advocated and were used in this case, but did not prevent the occlusion of the right iliac artery. Performing interventions using alternative routes such as axillary or carotid arteries, or using surgical cut-down, are potential options to consider on a case-to-case basis.

The Palmaz® stent and the Adventa® V12 stent are both expandable at 18 mm allowing balloon dilation to adult size dimensions.

Adventa stents are no more commercialized since the technical problem with reports of stent collapse in some patients.[12] Bentley® BeGraft aortic stent could potentially be used in the future in a similar situation. The 12 mm diameter premounted stent, requiring only a 9 F introducer, can be dilated up to a diameter of 20 mm. Also, the recently introduced Optimus™ stents (AndraTec GmbH), a balloon-expandable not premounted stent with a low profile and small sheath compatibility could be used in a similar situation. Those stents, dilatable up to 18 mm in diameter, can be mounted on variable delivery balloons, maximizing the treatment options.[13] According to the manufacturer, an Optimus™ L-stent mounted on an Altosa-XL noncompliant balloons of 12 mm (AndraTec) can be safely introduced in 9 F sheaths for noncovered stent and 10 F sheaths for covered Optimus™ L-stent. Using low-profile noncompliant balloon such as a Powerflex ™ Cordis 8 mm balloon (requiring a 5 F introducer) to mount the stent could even allow the use of 7 F sheaths for noncovered stent and 8 F sheaths for covered Optimus™ L-stent.

An important consideration is the proximity of the Adamkiewicz artery generally positioned between T8 and L2 vertebral segments.[14,15] The first angiography showed no vessel emerging from the tight aortic stenotic segment, allowing a safe dilatation and stent implantation at this level.

3.1 | Conclusion

Dilatation with a cutting balloon followed by stent implantation was an effective treatment option in this infant to relieve the obstruction in low thoracic aortic coarctation and prevent upper limb hypertension. The use of a 6 F sheath, required to allow the introduction of a Palmaz® PG1910P stents, mounted on a low profile 5 mm Opta® balloon, is possible in small patients. Covered Advanta® V12 requires a 9 F introducer, limiting its safe use to children above 10 kg. Both stents are dilatable to 18 mm, an adult size diameter. The long-term results are excellent with normal blood pressure, a patent aorta with no residual gradient on Doppler, and no arm–leg blood pressure difference at the age of 11 years.

AUTHOR CONTRIBUTIONS

Karlien Carbonez: Writing—original draft. **Joëlle Kefer**: Conceptualization, writing—review and editing. **Thierry Sluysmans**: Writing—original draft. **Stephane Moniotte**: Writing—review and editing.

CONFLICTS OF INTEREST

The authors declare no conflicts of interest.

TRANSPARENCY STATEMENT

T. S. affirms that this manuscript is an honest, accurate, and transparent account of the study being reported; that no important aspects of the study have been omitted.

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