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CASE REPORT

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Transcatheter management of adult aortic coarctation with "Railway" technique

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Abstract

Endografting for atretic coarctation is technically feasible to avoid the risks of open surgery. It requires a strategic and structured endovascular approach such as the "Railway" technique for safe and successful restructuring of complete aortic atresia and avoiding rupture or bleeding.

KEYWORDS

aortic coarctation, congenital cardiac defects, interventional, stent

1 | INTRODUCTION

We report the case of a 21-y-old man with complete atresia of the aorta who underwent percutaneous aortic intervention and successful recanalization of an atresia of the aorta using a "Railway" technique. A fully atretic aorta poses a challenge for a percutaneous solution and is often subject to open repair, considering the risk of perforation and technical failure. However, our case demonstrates that a strategic and structured approach is safe and successful to even tackle complete aortic atresia without complications such as rupture or bleeding.

Coarctation of the aorta (CoA) is a congenital stenotic malformation located in the proximal descending aorta distal to the origin of the left subclavian artery at the level of the arterial duct ("juxta-ductal"). CoA may vary in presentation, ranging from heart failure at neonatal age to upper body hypertension detected late in adolescence or adulthood.

CoA accounts for approximately 5%-8% of all congenital cardiovascular defects with a prevalence of 0.3-0.4 per 1000 live births.[1] In the majority of cases, CoA is associated with additional cardiovascular anomalies such as a bicuspid aortic valve (46%), patent ductus arteriosus (49%), mitral valve abnormalities, ventricular septal defect (49%), or additional obstructive lesions of the left heart.[2]

Timely surgical or endovascular repair of CoA is likely to abolish aortic obstruction and its sequelae, while untreated long-term hypertension and vascular dysfunction will result in an increased risk of permanent damage to organs exposed to hypertension such as heart and brain. Even after successful management long-term surveillance is advised since post-surgical aneurysm formation or restenosis after stenting has been observed.

2 | CASE REPORT

Here, we describe a 21-y-old asymptomatic male who was referred for management of hypertension; he was essentially fit with a BMI of 25 kg/m^2, on no medication but showing borderline left ventricular hypertrophy. On examination, his upper body blood pressure was 158/94 mm Hg while lower body blood pressure was 120/82 mm Hg. (Table 1) In his work-up, complete atresia of the aorta was identified as an

extreme form of coarctation with extensive collateralization both on CTA and MRI; the proximal aorta was completely interrupted with no luminal connection (Figure 1). Multidisciplinary team decision opted for an endovascular attempt to treat this patient, particularly in view of an extensive bleeding risk from the abundance of collaterals in case of a surgical approach.

The subsequent interventional procedure was performed under general anesthesia in a hybrid suite allowing conversion if needed; technical details and devices used for the procedure have been described previously.[3,4] After obtaining percutaneous access to the right femoral artery, an 8F introducer was placed to accommodate a pigtail catheter for an initial angiogram which documented a completely obstructed (atretic) aorta with no communication to the upper aorta (Figure 2A). A second angiogram from above was performed via a second pigtail catheter over a 5F introducer via right radial access to confirm complete atresia of the aorta at the typical level of adult coarctation about 1cm distal to a large left subclavian artery (Figure 2B); extensive collaterals were visualized with the injection of contrast into the aortic arch. As a special procedural step and different from nonatretic coarctation, an attempt was made to establish a connection across the atretic segment in antegrade rather than in retrograde fashion in order to avoid damaging and bleeding of a large collateral when going retrogradely (Figure 2A). Technically, a 5F right coronary artery catheter via the right radial access was navigated into the center of the occlusion of the upper aortic stump and gently rotated with moderate pressure. Once the catheter had settled in a stable position, it was possible to thread an extralong (260mm) coated Terumo® 0.035-inch hydrophilic wire through the atretic lesion and reach the distal aorta safely. (Figure 2C) The long Terumo® wire was then externalized via the right femoral artery by use of a snaring (Amplatz Goose Neck™, Medtronic®) procedure; the introducer in the right groin was then upsized to 14F and a 7x20 mm Powerflex™ balloon (Cordis®) was successfully advanced into the obstructed segment and dilated at 6 atmospheres. (Figure 2D,E) After preballooning and establishing a true anatomic connection between lower and upper aorta, the Terumo® wire was exchanged for an Amplatz super-stiff™ wire (Boston Science©). Once the Amplatz super-stiff™ wire was parked in the descending aorta, a 14F Performer™ Mullins sheath (Cook® Medical) was advanced across the recanalized aortic segment. As a next step, a 45 × 22 mm bare CP (Cheatem-Platinum) stent® (B.

TABLE 1 Hemodynamic information before and after intervention to complete atresia

	Before intervention	After intervention
Blood pressure (mm Hg)		
Arm	158/94	105/70
Leg	120/82	105/60
Ankle-brachial index	0.76	1
Coarctation gradient (mm Hg)	38	0

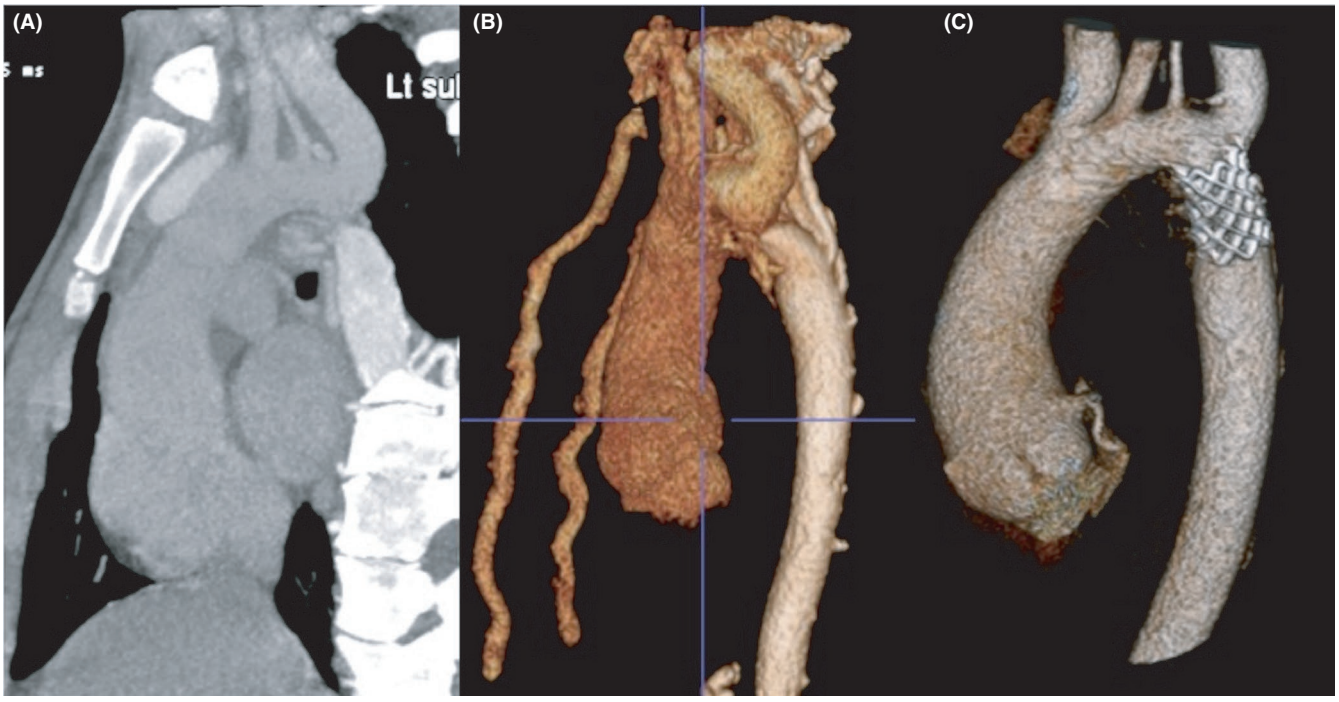

FIGURE 1 CTA shows complete atresia (asterisk) between the upper and lower thoracic aorta in a sagittal view (A) and 3D reconstruction (B) with no flow communication or minimal stenotic diameter; (C) 7 months follow-up CT shows the stent lumen widely patent, a homogeneous diameter of the aorta and no obstruction of the left subclavian artery

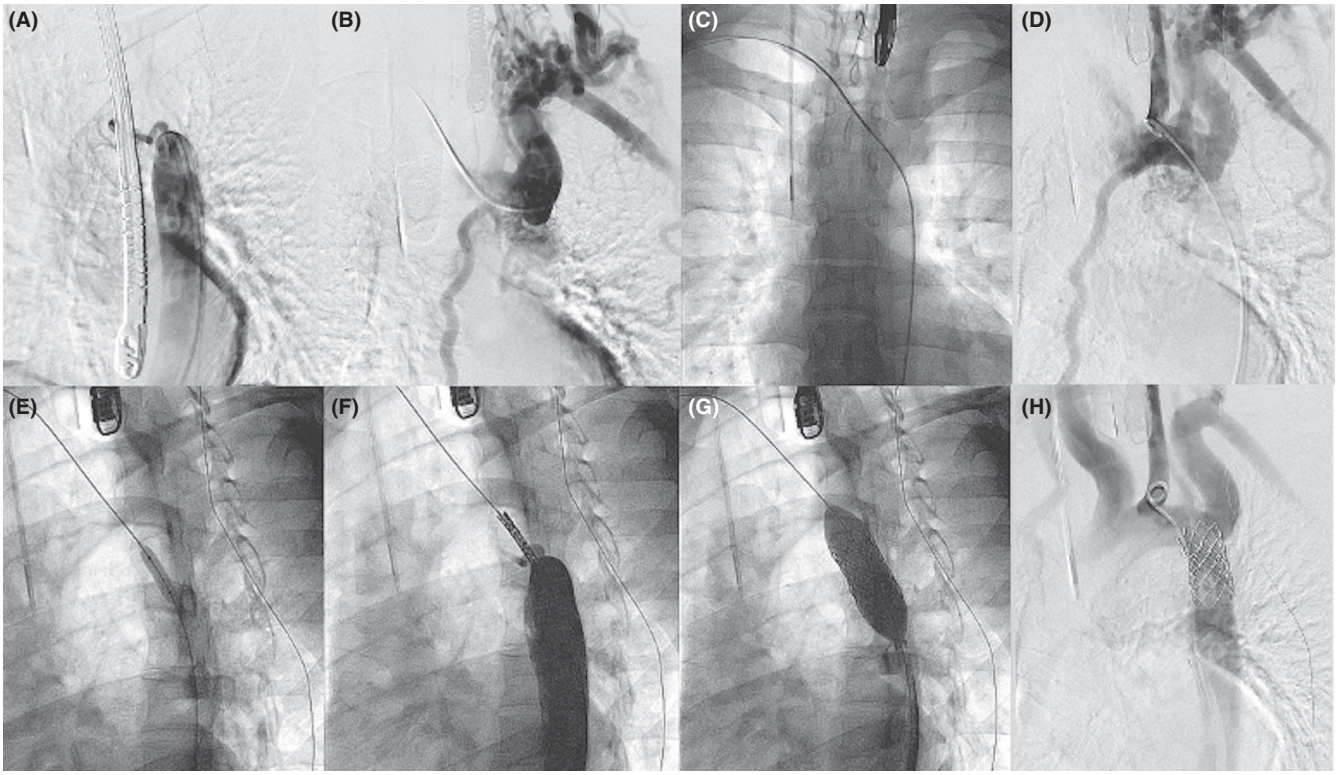

FIGURE 2 Contrast angiogram from below (A) and above (B) showing no communication between upper and lower aorta; (C) a hydrophilic Terumo® wire was successfully passed through the obstructed segment; (D) angiogram confirming the pigtail catheter across the obstructed segment from below; (E) a 7 × 20 mm Powerflex™ balloon was used to predilate the atresia; (F) CP stent positioned at the target segment; (G) CP stent deployed with a BIB balloon; (H) final angiogram showing a satisfactory result without any complication

Braun Medical) was crimped onto a BIB® balloon (B. Braun Medical) and gently advanced through a 14F Performer™ Mullins sheath to be positioned exactly at the level of the recanalized aorta. (Figure 2F, 2G) With sequential ballooning of the inner and outer balloon, the CP stent was precisely placed with no residual waste and expanded to the diameter of the thoracic aorta leaving the LSA (left subclavian artery) unobstructed. A completion aortogram showed the fully open stent precisely on target with no obstruction of the left subclavian artery and no extravasation of contrast (Figure 2H); these findings were confirmed by simultaneous transoesophageal ultrasound (TOE) imaging before terminating the procedure and removal of all access sheaths. The patient was extubated and, on table, monitored for 4 hours prior to moving to a step-down unit and discharged 3 days later with a completely abolished transcoarctation. (Figure 1 and Table 1) A follow-up CT angiogram was scheduled 6 months later.

3 | DISCUSSION

Coarctation repair is recommended in adult patients regardless of symptoms in presence of either radial-femoral arterial pulse delay, or catheter-derived peak-to-peak gradient of >20 mm Hg at rest of >30 mm Hg with exercise, and in difficult to manage hypertension.[5] Moreover, relief of aortic coarctation may even be indicated if radiologic evidence shows significant collateral flow, and a low gradient, with the intention to decrease cardiac afterload and deleterious long-term consequences such as progressive left ventricle hypertrophy, or concomitant conditions like aortic aneurysm formation or premature coronary or carotid atherosclerosis. The benefit and appropriate threshold for an intervention are less clear in patients with moderate coarctation who exhibit hypertension with exercise only.

Cardiovascular specialists generally agree that percutaneous angioplasty, with stent or stent-graft implantation, is the preferred nonsurgical management option for both congenital and recurrent aortic coarctation.[3,6] While percutaneous treatment strategies for adult coarctation have essentially replaced open surgery,[6,7] a fully atretic aorta poses a challenge for any percutaneous solution [8] and is in general subject to open repair in view of the risk of perforation and technical failure. However, our case demonstrates that a strategic and structured approach is safe and successful to even tackle complete aortic atresia without complications such as rupture or bleeding. The strategy encompassed 3 elements to ensure a safe procedure. First, recanalization (eg, wire perforation at the atretic segment) was performed in antegrade fashion from above with gentle pressure using a 0.035-inch hydrophilic

wire; even a perforation of the aortic wall with such a wire would not cause serious bleeding issues. Second, once a connection to the lower aorta was established and confirmed on both TOE and fluoroscopy, gentle predilation was performed from below to generate space for the subsequent CP stent and to get a feel for the distensibility of the atretic aorta. Finally, a bare CP stent was gently inflated in steps to minimize the risk for any bleeding in case the expanded tissue would tear. For such a scenario, a further back-up strategy was in place by having a suitable stent-graft device ready to be used and by performing the procedure in a hybrid suite rigged for optional conversion. Thus, we think with this set-up and match plan the procedure was as safe as in the setting of nonatretic CoA. The efficacy and safety profile of stenting in classic (nonatretic) CoA are known to be excellent; for instance, the National Cardiovascular Data Registry reports that of 671 patients having angioplasty and stenting of coarctation, 351 patients (52.4%) had a resting gradient of ≥25 mm Hg, a further 143 (21.4%) experienced hypertension, 108 patients (16.1%) had angiographic evidence of coarctation and the final percentile (10.2%), or 68 patents developed left ventricular failure.[9] While the rate of success is high, it is important to note that 73.6% of stenting cases were patients above 18 years. Minor complications such as bleeding were observed in 17% of patients and a further 1.2% of patients experienced major complications requiring ancillary procedures; one death was the result of vascular complications. A separate study by the CCISC (Congenital Cardiovascular Interventional Study Consortium) compared stent implantation across a number of procedures (n = 27) with angioplasty alone (n = 61) and surgery (n = 72) for coarctation. [10] No major events were associated with interventional procedures in coarctation; thus, stenting is the preferred treatment for patients above 25 kg. Although there are no published data yet for completely atretic aortas, the risk of stent migration, stent collapse or fracture, or even dissection is unlikely to be higher than in classic coarctation.

Covered or uncovered balloon-expandable devices for the treatment of adult coarctation were compared in a randomized fashion with a follow-up of 31 ± 19 months; a reintervention was required in just one case of an uncovered stent. [6] Both covered and uncovered stents provide reliable hemodynamic improvement and procedural safety regardless of coarctation severity; similarly, both configurations of the CP stent were recommended in postductal coarctation,[6] while its suitability for other anatomic variants of coarctation may vary. Due to the complex nature of aortic pathology, however, each case should be assessed individually. Nevertheless, stent technology is improving and promising future advances in stent technology including biodegradable platforms may even offer a sustainable solution with nonsurgical management in infants and children with coarctation and allowing them a normal growth.

4 | CONCLUSION

A strategic and structured endovascular approach such as the artery "Railway" technique is safe and successful to even tackle complete aortic atresia without complications such as rupture or bleeding.

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Published with written consent of the patient.

CONFLICT OF INTEREST

The author(s) declared no conflicts of interest with respect to the research, authorship, and/or publication of this article.

AUTHOR CONTRIBUTIONS

MR and CAN: involved in conceptualization. XY and AM: involved in data curation. XY and CAN: involved in writing.

ETHICAL STATEMENT

Published with written consent of the patient.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

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