

Complicated coarctation repair: The importance of three-dimensional cross-sectional imaging in late postoperative assessment

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ABSTRACT

Coarctation of the aorta (CoA) represents 5%–8% of congenital heart disease patients and is one of the most common causes of neonatal surgical intervention. These patients require close lifelong follow-up due to frequent long-term complications. Although transthoracic echocardiography is the first-line technique for its diagnosis and follow-up, cross-sectional imaging with cardiovascular magnetic resonance (CMR) gives excellent anatomical and functional information, especially in complex CoA. We present the case of a 17-year-old patient who underwent complicated neonatal CoA repair and demonstrate how CMR and thorough operative records helped to define the exact anatomy of repair many years after surgery. Furthermore, we conclude that keeping surgical drawings in the patient records can be of great importance, especially in complicated cases.

Keywords: Aortic coarctation, cardiovascular magnetic resonance imaging, cross-sectional imaging, re-coarctation

INTRODUCTION

We present the case of a 17-year-old adolescent who was diagnosed with coarctation of the aorta (CoA), hypoplastic aortic arch, and bicuspid aortic valve at day 7 of life. Neonatal surgery was difficult leaving the patient with an unusual and complex aortic anatomy and complicated long-term follow-up. Our report demonstrates not only the importance of thorough operative records and drawings, but also the availability of three-dimensional (3D) cross-sectional imaging data for detailed aortic anatomy description.

CASE REPORT

Neonatal surgical CoA repair was performed on a 6-day-old male infant and was intraoperatively complicated by difficulty in establishing sufficient distal aortic blood flow. Initially, an extended end-to-end

anastomosis via left posterolateral thoracotomy was performed, but there was no distal aortic pulsation following the procedure. The surgeon then proceeded to create an additional end-to-side anastomosis between the left common carotid artery (LCA) and the divided left subclavian artery (LSA). However, there was still a persistent aortic gradient without distal aortic pulsation. Finally, a 6-mm extra-anatomic GORE-TEX® graft was implanted, extending from the mid-aortic arch to the proximal descending aorta, beyond the area of CoA repair with good result.

At the age of 5 years, the patient presented with an increased Doppler gradient across the aortic isthmus. He subsequently underwent successful balloon dilatation of what was considered to be the native aortic isthmus but which, retrospectively, was the

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anastomosis between the LCA and the LSA. Interpretation of the cardiovascular magnetic resonance (CMR) images acquired at that time was challenging as the scan was performed without general anesthesia or sedation, resulting in less good image quality, and operation notes were not available. Furthermore, contrast-enhanced or noncontrast-enhanced magnetic resonance angiograms (MRA) were not acquired due to anxiety of the patient. Another routine CMR scan performed at the age of 12 years described a narrow native aortic pathway, which in retrospect described the extra-anatomic graft and the LCA and proximal LSA anastomosis. However, the image analysis was limited because 3D angiographic data were not possible to obtain again due to anxiety. No further specific procedures were done at that time as there were no alerting clinical or imaging signs of re-coarctation.

When the patient was 14 years of age, he presented again but this time with clinical signs of re-coarctation. The patient underwent cardiac catheterization with successful endovascular stent implantation of the LCA and proximal LSA anastomosis [Figure 1, ++]. Visualizing the re-coarctation and the anatomy of the area in the catheter lab was technically challenging, but it was thought by the operator that there was probably a multilevel stenosis with the main narrowing at the isthmus level that led to the decision of immediate stenting (length of the stent 34 mm) followed by re-dilatation of the stent with 14 mm × 20 mm balloon. The result after stent implantation was good with only mild residual gradient (10–11 mmHg) due to transverse arch hypoplasia.

At the latest follow-up, the now 17-year-old patient was again found to have a raised right arm arterial blood pressure and an arm-leg blood pressure gradient of >20 mmHg. Doppler echocardiogram demonstrated

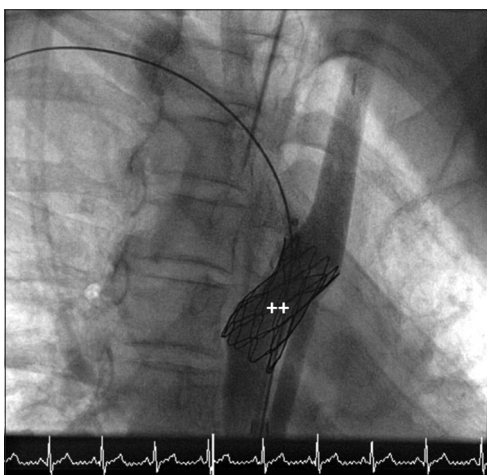


Figure 1: Catheter angiogram with stent implantation of the left common carotid artery and proximal left subclavian artery anastomosis (++)

an increased gradient across the stented aortic area, and pulse-wave Doppler interrogation of the abdominal aorta showed an abnormal abdominal aortic flow pattern with dampening of the flow signal indicating lower body ischemia. Otherwise, the patient was clinically well and asymptomatic without additional cardiovascular risk factors. As part of the workup, a repeat CMR scan with acquisition of a noncontrast 3D steady-state free precession (3D SSFP) MRA was undertaken. During image analysis, review of the surgical notes and drawings, now available, was particularly important to define the detailed aortic anatomy [Figure 2]. CMR showed a 6-mm extra-anatomic graft from the distal aortic arch to the descending aorta beyond the coarctation stent. The tortuous LCA and proximal LSA anastomosis (previously thought to be the native stenotic isthmus) was stented [Figure 2b and c, ++]. Even though spin echo sequences are typically advantageous compared to SSFP cine imaging to image-stented areas as they are less susceptible to artifacts caused by metallic implants,^[1] in our patient, they were of suboptimal quality due to extensive motion artifacts. Phase-contrast flow imaging showed that there was no blood flow across the native isthmus, suggesting complete obstruction of the native aortic pathway although the assessment was slightly limited because of the stent artifact. It also did not indicate significant collateral flow. Analysis of combined cine and 3D isotropic balanced SSFP (b-SSFP) imaging showed overall that the patient had significant obstruction into the descending aorta with restriction of flow both through the small 6-mm interposition graft and also through the anastomosis between the LCA and proximal LSA. A successful image analysis and interpretation, however, was only possible with the help of surgical notes and drawings from the neonatal surgical CoA repair.

The case was discussed in the joint cardiology and surgical conference multidisciplinary meeting at our institution and the patient underwent further interventional cardiac catheterization with endovascular stenting (length of the stent 45 mm) of both sites of stenosis, i.e., the distal aortic arch and the proximal part of the LCA-to-LSA anastomosis. The angiographic and hemodynamic result was good with a residual gradient of <10 mmHg.

DISCUSSION

CoA is a lesion requiring lifelong follow-up due to potential complications.^[1,2] Nowadays, CoA is commonly detected early after birth, and patients often undergo surgical repair in the neonatal period. The choice of surgical therapy depends on the underlying anatomy, with extended end-to-end anastomosis being the preferred surgical method at present.^[3] Although surgery for CoA is a standard procedure, it can be complicated

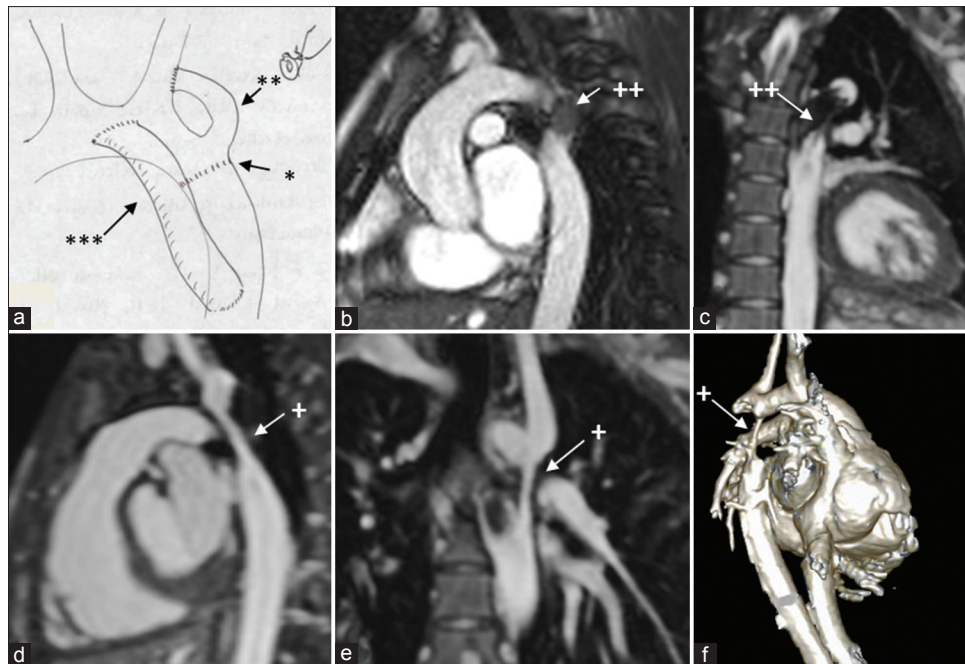


Figure 2: (a) The surgeon's original drawing demonstrating the end-to-end anastomosis (*), left common carotid artery and proximal left subclavian artery anastomosis (**), and extra-anatomic graft (***) techniques utilized for coarctation repair. (b and c) Two-dimensional steady-state free precession cardiovascular magnetic resonance images showing the modified and stented subclavian flap (**). (d-f) Multiplanar reconstruction and three-dimensional reconstruction illustrating the 6-mm extra-anatomic graft (+) from the distal aortic arch to the descending aorta beyond the coarctation area

by several factors, making further interventions necessary as demonstrated in our patient. Modern guidelines, therefore, recommend baseline and follow-up imaging using CMR or computed tomography (CT) for visualization of the aorta and early detection of long-term complications.^[2,4,5] In our patient, CMR imaging was performed several times; however, at the time of the first two scans, imaging was incomplete due to patient anxiety. Furthermore, surgical drawings as well as notes were not available, which made the interpretation of the images extremely difficult. Cardiac catheterization is still used at many centers for evaluation and typically indicated for interventional treatment of re-coarctation.^[3] In complex patients, 3D images from rotational angiography can also be of help for the guidance of complex diagnostic and interventional catheterization procedures but was not performed in our patient.^[6]

This report demonstrates that the combination of 3D cross-sectional imaging, phase-contrast flow imaging, and focused cine imaging based on thorough operative records and visual drawing of the interventions during operation is essential to provide anatomical clarity sometimes lacking from mere knowledge of other imaging modalities.

We recommend keeping surgical drawings in the patient's records along with a detailed operative letter to assist planning and interpretation of the future regular follow-up imaging needed in this population. 3D multiplanar imaging should be unequivocally included in the follow-up of all post-CoA

repair cases, especially for those complicated by either complex/serial interventions or clinical signs of re-coarctation. There is a lot of debate on whether CT or CMR imaging should be preferred in stented cases. Artifact and signal loss from stents *in situ* in magnetic resonance imaging can be surpassed by multiplanar spin echo or "black blood" imaging and nowadays also with isotropic 3D volume-balanced b-SSFP sequences which will also allow for very detailed evaluation of anatomy and stent position and enable complication detection and further management planning.

Advanced visualization unquestionably aids cardiac imaging; however, it is the profound knowledge of the patient's clinical background and surgical history that leads to deep understanding of the clinical problem, an absolute prerequisite of good medical practice and successful management planning.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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