

# Technical considerations of coarctation stenting in double-barreled aorta – A persistent fifth arch mimic

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## ABSTRACT

The double-barreled aorta connecting the ascending aorta and descending aorta caudal to the normal fourth aortic arch has fascinated the interests of cardiac morphologists for over a century. This condition is commonly associated with coarctation. While the controversies surrounding the embryology of the double-barreled aorta have settled down, we present a case-based illustration of the technical aspects of coarctation stenting peculiar to this condition.

**Keywords:** Aortic arch, coarctation of aorta, congenital heart disease, double barreled aorta, fifth arch

## INTRODUCTION

Abnormalities of development of the aortic arch have fascinated the interests of cardiac morphologists for over a century. Controversies around the explanations of the fifth pharyngeal arch have never died down for want of consistent temporal proof of its persistence in human embryological studies. Since the upper end of the pulmonary arch is the homologue of the distal portion of the fifth pharyngeal arch,<sup>[1]</sup> an extrapericardial vascular channel that connects the ascending aorta to the pulmonary artery or adjacent dorsal aorta is probably the closest to the putative definition of a persistent fifth arch.<sup>[2]</sup> The double-barreled aorta connecting the ascending aorta and descending aorta caudal to the normal “fourth” aortic arch is probably better considered a “fifth arch mimic” rather than a persistent fifth arch as some evidence points to island formations immediately caudal to the fourth arch which gain connections to the descending aorta.<sup>[3,4]</sup> This condition is often associated with coarctation of aorta. We present a case-based

illustration of the technical aspects of percutaneous stent implantation in this condition.

## CASE REPORT

A 19-year-old boy was referred to our center for systemic hypertension detected during an episode of viral fever. On examination, his heart rate was 70/min and the lower limb pulses were weak. The right upper limb blood pressure was 186/110 mmHg with a differential blood pressure gradient of 60 mmHg between the upper and lower limbs. A grade two mid-systolic murmur was noted in the left second intercostal space. Chest X-ray showed bilateral rib notching from the third to sixth ribs. The electrocardiogram showed left ventricular hypertrophy and normal QRS axis.

Transthoracic echocardiography confirmed severe coarctation of aorta and left-sided aortic arch. Imaging from the suprasternal window demonstrated a double-barreled aortic configuration [Figure 1]. The superior arch gave rise to all three neck vessels.

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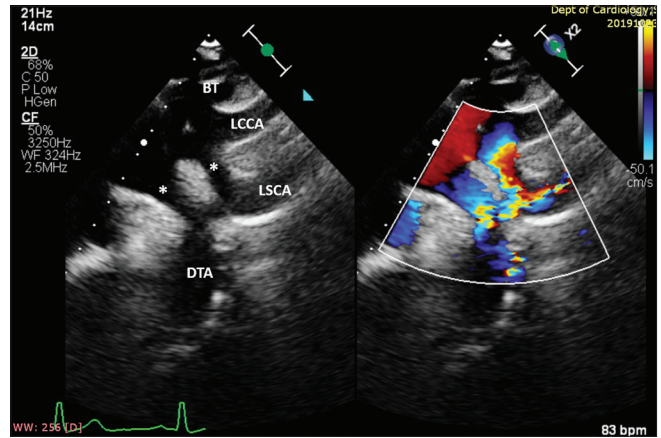
A caudally located arch connected the distal ascending aorta to the aortic isthmus. The arterial duct was not patent. Discrete isthmic narrowing at the confluence of both aortic arches amounted to severe coarctation with a peak gradient of 50 mmHg and diastolic tailing. The left ventricle was hypertrophied with good systolic function.

Cardiac computed tomography confirmed that the caudal arch connected to the ascending aorta just proximal to the origin of the right brachiocephalic trunk. The superior arch had critical narrowing immediately beyond the origin of the left subclavian artery. Stenosis at the confluence of both arches assumed a bifurcation lesion configuration [Figure 2]. Both the mid-transverse arch and the inferior arch measured 15 mm, and the descending thoracic aorta measured 17 mm at the diaphragmatic level. The narrowest portion at the junction of the inferior arch to the descending aorta measured 6 mm. Collateral vessels were identified between the branches of the subclavian artery and intercostal arteries.

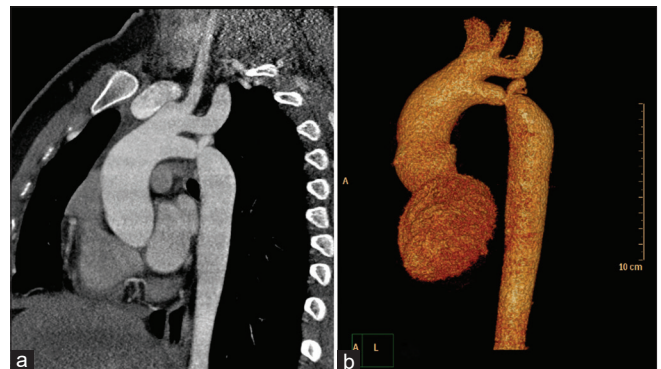
Cardiac catheterization was performed under general anesthesia. A 30 mmHg pullback gradient was obtained across the coarctation. Aortic angiography confirmed that the lower arch was more in line with the descending aorta while the superior arch presented an acute angle at the confluence and gave rise to the neck vessels [Figure 3a and Video 1]. Hence, it was decided to stent the coarctation into the inferior arch. An Amplatzer superstiff wire was positioned in the proximal ascending aorta through the inferior arch. A 35 mm uncovered Andra XL (Andramed, Reutlingen, Germany) stent was mounted on a 16 mm × 40 mm Andra balloon XL (Andramed, Reutlingen, Germany) and delivered over a 12F Mullins sheath. The stent was positioned into the inferior arch and expanded to 16 mm [Figure 3b]. The distal edge of the stent was flared with an 18 mm × 30 mm Tyshak II (NuMed Cornwall, Canada) balloon. The pressure gradient after stenting was 6 mmHg. Postprocedure angiogram confirmed

satisfactory stent position [Figure 3c and Video 2] and no complications. The flow into the neck vessels was normal.

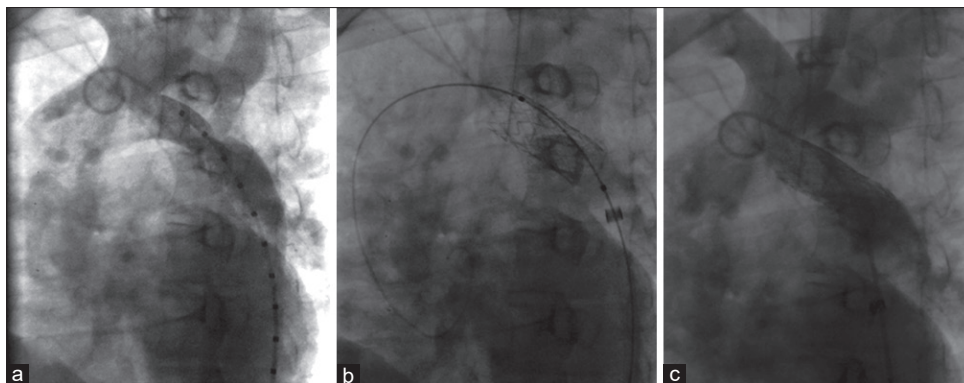
Systemic hypertension resolved fully on follow-up with no differential blood pressure recording in any limb. The flow in the arch vessels was normal. He is doing well at latest follow-up at 9 months.



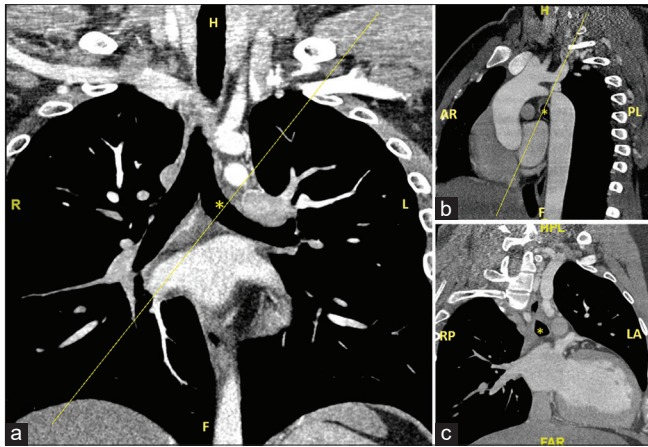
**Figure 1: Transthoracic echocardiography from the suprasternal projection showing the double-barreled aorta (asterisks) with coarctation. BT: Brachiocephalic trunk, DTA: Descending thoracic aorta, LCCA: Left common carotid artery, LSCA: Left subclavian artery**



**Figure 2: CT aortogram (a) and 3D reconstruction (b) showing double-barreled aorta with coarctation at the confluence of the two arches conforming to a bifurcation lesion**



**Figure 3: (a) is the aortic angiogram in the left anterior oblique 50° view showing the double-barreled aorta with coarctation. The stent was deployed into the inferior arch (b). (c) is the postprocedure angiogram**



**Figure 4:** CT aortogram in coronal (a), oblique sagittal (b) and right anterior oblique (c) sections showing the relationship of the inferior arch of double-barreled aorta to the left bronchus (yellow asterisk). AR: Anterior right, H: Head, HPL: Head posterior lateral, F: Foot, FAR: Foot anterior right, L: Left, LA: Left anterior, PL: Posterior lateral, R: Right, RP: Right posterior

## DISCUSSION

Double-barreled aorta (or double-lumen aortic arch) is considered as a type 1 persistent fifth arch.<sup>[5]</sup> However, it does not fulfill all the criteria of the fifth arch artery to be unequivocally considered its vestige. While recent work by Gupta *et al.* seems to have laid rest to the embryological origins of this entity,<sup>[2,3]</sup> this condition poses specific challenges in percutaneous coarctation stenting. The confluence of both the arches is the most common site of coarctation and both the inserting limbs are often involved resulting in a true bifurcation lesion.<sup>[6]</sup>

The salient principle in percutaneous stenting of double-barreled aorta is identification of the dominant vessel that is stented and the smaller one is left as a side branch.<sup>[7]</sup> Serial balloon angioplasty and surgery is an option for patients presenting in the neonatal period.<sup>[8]</sup>

In a double-barreled aorta, while the superior arch gives rise to the neck vessels, the inferior arch is nonbranching and may be the dominant vessel, as in the index case. This offers the options of choosing either an uncovered or covered stent without side branch or neck vessel compromise despite the jailing involved. The inferior arch often provides a more favorable angle for stenting. The latter's relative proximity to the left bronchus should also be considered during stent placement (10 mm in our case) [Figure 4]. The native fourth aortic arch also tends to have more severe stenosis immediately beyond the left subclavian artery due to isthmic ductal tissue.

While the diameter of the inferior arch dictates the balloon size, the absence of branch vessels from it also permits considerable leeway in stent length selection. A balloon-in-balloon offers more precision in stent positioning.<sup>[7]</sup> The incremental benefits of a covered stent over a bare metal

stent in this scenario is limited to tight coarctation in older children and adults as a safeguard for aortic wall injury.<sup>[9]</sup>

## CONCLUSIONS

A double-barreled aorta with coarctation can be safely treated with bare metal stent implantation into the lower arch. Consideration of the anatomic peculiarities is associated with a good procedural result.

### Declaration of patient consent

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

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Nil.

### Conflicts of interest

There are no conflicts of interest.

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