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MINI-FOCUS ISSUE: TRANSCATHETER INTERVENTIONS

ADVANCED

CASE REPORT: CLINICAL CASE

Rerouting of Cerebral Circulation

Extensive Transcatheter Aortic Arch Revision

R. Allen Ligon, MD, Denver Sallee, MD, Sassan Hashemi, MD, Clifford M. Hawkins, MD, Christopher J. Petit, MD

ABSTRACT

We describe an adolescent with long-standing atresia of the head/neck arteries and severe aortic coarctation. Because of progressive symptoms, a series of interventions was undertaken to provide direct aorta-to-carotid artery flow and coarctation treatment. This case highlights the unusual physiological features associated with atresia of all head and neck arteries. (**Level of Difficulty: Advanced.**) (J Am Coll Cardiol Case Rep 2020;2:855–9) © 2020 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 13-year-old boy was electively admitted for transcatheter vessel recanalization/intervention.

PAST MEDICAL HISTORY

He had a large ischemic stroke in infancy presented with arteriopathy of unclear etiology (aortopathy panel and whole-exome sequencing demonstrated no pathogenic variants). The stroke in infancy had resulted in cortical blindness and right-sided hemiplegia. At the time of the stroke, work-up revealed extensive aortic arch anomalies, including atresia of the right innominate artery (RIA), left common carotid artery, and left subclavian artery.

LEARNING OBJECTIVES

- To appreciate that long-standing arterial atresia in children can be recanalized and stented, even within the head and neck vasculature.
- To understand that coarctation of the aorta can present with an abnormally high echocardiographic gradient when there is a paucity of collaterals present.
- To recognize that with atresia of head/neck vessels, there can be flow reversal in the collaterals that supply the head vasculature.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis included genetic/familial thoracic aortic aneurysm and dissection (TAAD), nonsyndromic TAAD, genetic versus nonsyndromic aortopathy, prenatal ischemic insult, or aortic coarctation.

INVESTIGATIONS

Subsequent computed tomographic angiography (CTA) demonstrated reconstitution of the distal carotid arteries and subclavian arteries, fed by networks of small arteries from the descending aorta (**Figures 1A and 1B**, Video 1). Over the ensuing years, the child thrived, attending special education and generally remaining clinically stable. Over the past year,

From Children's Healthcare of Atlanta, Department of Pediatrics, Emory University School of Medicine, Atlanta, Georgia. The authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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ABBREVIATIONS AND ACRONYMS

CTA = computed tomographic angiography

RCA = right carotid artery

RIA = right innominate artery

TAAD = thoracic aortic aneurysm and dissection

however, he began to experience lightheadedness and confusion, particularly during exertion or challenging homework. Recent echocardiography noted progressive left ventricular hypertrophy and an aortic coarctation peak velocity of 8 m/s by Doppler (estimated mean gradient: 143 mm Hg) (Figure 2).

In the past, his family had been reluctant to undertake any intervention out of fear of potential adverse sequelae. However, at this point, given the patient's symptoms and the findings on imaging, the consensus among multiple specialists was to attempt to recanalize the patient's RIA. Vascular and cardiovascular teams both evaluated the patient but ultimately believed the transcatheter approach would be optimal.

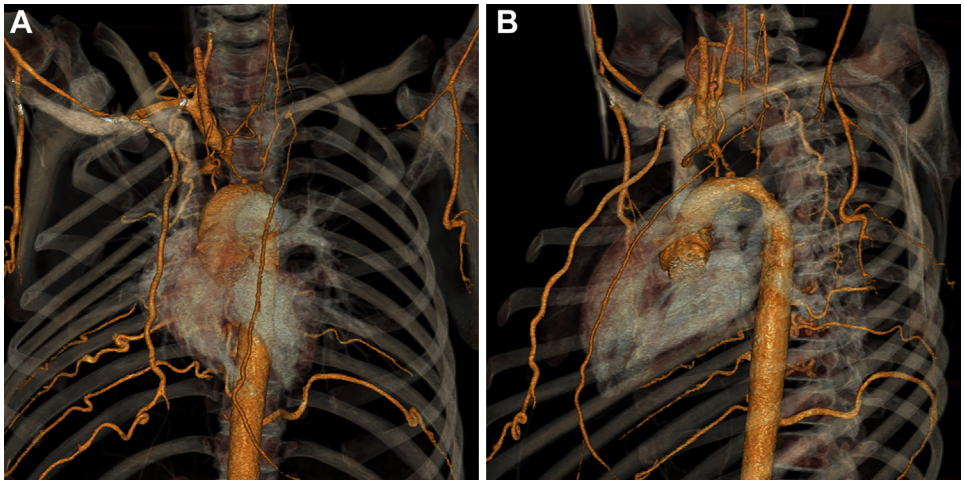
MANAGEMENT

On the day of the initial intervention, the patient underwent a right and left heart catheterization, which revealed a left ventricular pressure of 170/14 mm Hg, with a gradient of 90 mm Hg across the aortic arch. Percutaneous right carotid artery (RCA) access was obtained by using ultrasound guidance, and the pressure in the RCA was 55/49 mm Hg. Using a chronic thrombotic occlusion wire, the RIA was recanalized and carefully angioplastied to a 5-mm diameter (Figures 3A and 3B). The vessel remained severely stenotic and therefore was stented with a pair of Onyx drug-eluting stents (Medtronic, Santa Rosa, California) (Figure 2A). Afterward, there was no difference in pressure between the RCA and the descending aorta (72/52 mm Hg, respectively), but there was a gradient of 90 mm Hg across the aortic arch.

Because of concerns of disrupting the freshly recanalized RIA, the patient was brought back 3 months later for arch intervention. At this second intervention, he underwent repeat angioplasty of the stented RIA up to 7 mm with an ultra-high-pressure Dorado balloon (24 atm, Bard, Tempe, Arizona). At this time, aortic coarctation stenting was performed. Because the gradient began just at the level of the stented RIA, a series of stents was required (26 mm, 36 mm, and 36 mm ev3 IntraStent Max Large Diameter Biliary Stent; Medtronic, Santa Rosa, California) to treat the entirety of the coarctation to relieve the gradient. Because the existing Onyx stent was jailed and disrupted by the aortic arch stents, the RIA was re-entered and restented proximally with a premounted stent dilated up to 8 mm in diameter (Figures 4A and 4B). At the conclusion of this second procedure, the arch gradient was only 2 mm Hg, and a gradient from the aortic arch to RIA of 12 mm Hg was measured. Pressure in the left ventricle dropped to 105/9 mm Hg.

The patient was treated with clopidogrel and aspirin for 6 weeks following the second procedure. A follow-up CTA 6 months later demonstrates an unobstructed RIA and aortic arch (Figures 5A and 5B).

FIGURE 1 Pre-Procedural Computed Tomographic Angiography



Present were **(A)** an atretic right innominate artery with limited collateralization that filled in a retrograde fashion, from lower extremity/chest wall to the cerebral vasculature, and **(B)** a notable aortic coarctation.

The patient has improved significantly, with improved energy. He is now able to attend school and no longer has dizziness or fatigue.

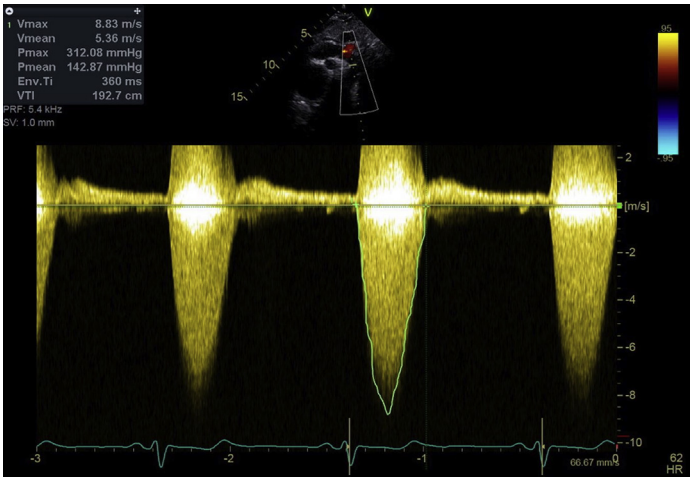
DISCUSSION

To our knowledge, our case is the first to demonstrate effective transcatheter therapy to treat atresia of the innominate artery in the setting of severe coarctation of the aorta. Although there are reports of adults undergoing carotid artery recanalization following acquired atherosclerotic atresia, this has not been demonstrated in children without atherosclerosis as the inciting pathology (1,2). Our patient underwent transcatheter recanalization of a chronically occluded innominate artery via a percutaneous carotid artery approach (3,4).

A unique aspect of the patient presentation was the presence of retrograde flow (from lower extremity and thoracic cavity up to the cerebral circulation) within chest wall collaterals present on CTA. In combination with the severe coarctation of the aorta, this led to a fascinating physiology. The presence of severe coarctation of the aorta would normally result in carotid arterial hypertension; however, with atresia of all head and neck vessels, cerebral blood flow in our patient was maintained solely through intercostal arteries supplying the internal mammary arteries. From the right more so than left internal mammary artery, flow ascended to the subclavian arteries, supplying the respective carotid arteries. Relief of severe coarctation in this setting would be expected to enhance the pressure available to perfuse the carotid arteries—even in the absence of RIA recanalization.

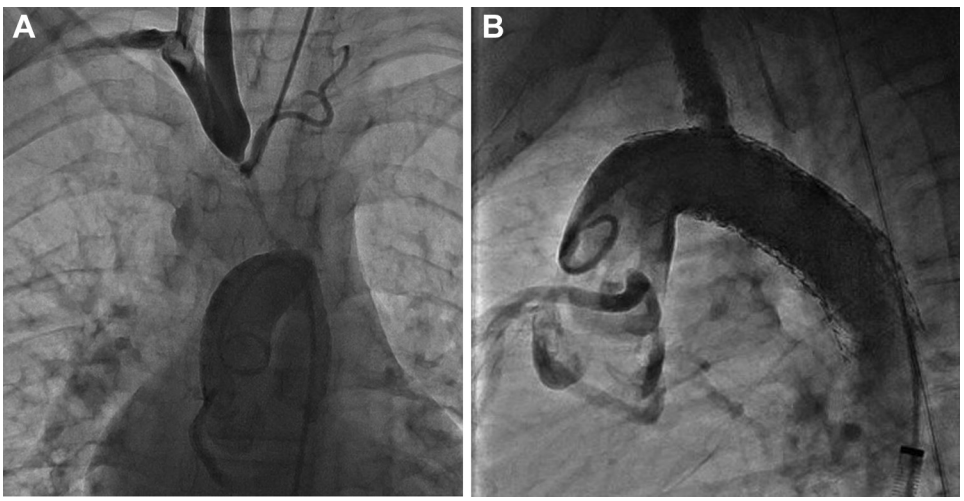
Of further interest was the outstanding pressure gradient across the aortic arch in our patient. Even in

FIGURE 2 Doppler Ultrasonography of the Aortic Arch



Across the aortic isthmus, there was a flow acceleration gradient of 8.8 m/s.

FIGURE 3 Intraprocedural Angiography



(A) An aortogram in the anterior-posterior dimension demonstrates an atretic right innominate artery (RIA), with no direct communication from aorta to the RIA. **(B)** Final angiography after coarctation and RIA angioplasty demonstrates no residual obstruction across the arch and an unobstructed RIA.

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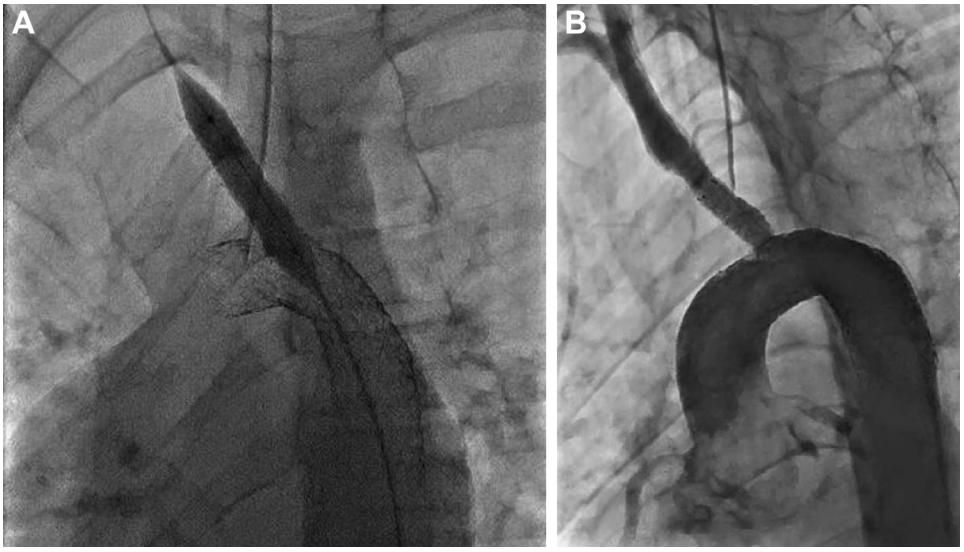
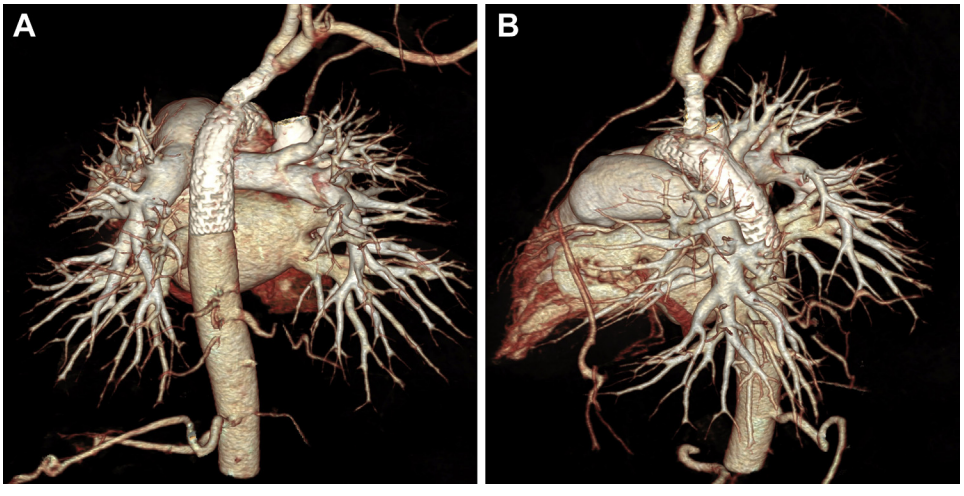
FIGURE 4 Intraprocedural Angiography



After vessel recanalization and stenting, **(A)** a left anterior oblique projection of an aortogram shows improved antegrade flow to the atretic right innominate artery and **(B)** no contrast extravasation.

the setting of near atresia of the aortic isthmus, systemic arterial gradients of this degree are not encountered. However, in all other cases of coarctation, a portion of the cardiac output will necessarily be allocated to the upper body, and this proportion of cardiac output is in fact enhanced by the presence of upper- to lower-body systemic arterial collateralization. Hence, the isthmus in the typical coarctation may be expected to handle a small proportion (0% to 25%) of the total cardiac output. In our patient, the unusual aortic arch anatomy resulted in 100% of cardiac output passing through the coarctation

FIGURE 5 Follow-Up Computed Tomographic Angiography



Following 2 transcatheter interventions, including coarctation stenting, imaging 6 months later demonstrates no evidence for obstruction to the areas **(A)** of aortic coarctation or **(B)** of the innominate artery as it enters the aorta.

segment. Without head and neck vasculature to provide a pop-off to circulation, the entire cardiac output passed through the aortic arch.

FOLLOW-UP

Following these interventions, the patient experienced considerable improvement of clinical status—no further events suggestive of cerebral hypoperfusion and a return to activities of daily living without difficulty.

CONCLUSIONS

The consideration of recanalization of the RIA and coarctation intervention in this child warranted careful planning. The operators and treatment team weighed the risks of development of carotid artery hypertension, thromboembolic complications following recanalization, and rebound hypertension. Although cardiac and vascular surgical teams demurred from participating in the treatment, we relied on a team of experts including interventional radiology and vascular surgery, as well as cardiology. In our patient, this staged approach to reconstruction of the aortic arch with recanalization of the RIA was successful in relieving severe left ventricular hypertension and provided enhanced carotid arterial flow.

ADDRESS FOR CORRESPONDENCE: Dr. R. Allen Ligon, Children's Healthcare of Atlanta, Emory University School of Medicine, Department of Pediatrics, 1405 East Clifton Road, Atlanta, Georgia 30322. E-mail: allenligon@gmail.com.

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KEY WORDS cardiac catheterization, carotid artery, pediatric cardiology, vascular recanalization

APPENDIX For a supplemental video, please see the online version of this paper.