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CONGENITAL HEART DISEASE

IMAGING VIGNETTE: CLINICAL VIGNETTE

Recurrent Pediatric Respiratory Distress From a Challenging Vascular Anomaly

The Uncrossing Operation for Circumflex Aorta

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ABSTRACT

The authors report a case of circumflex aorta causing persistent respiratory distress in a 9-year-old boy who had previously undergone vascular ring division and multiple aortopexies. The uncrossing operation was performed, with symptomatic relief of both posterior and right-sided tracheal compression from the transverse aorta and right aortic arch, respectively. (J Am Coll Cardiol Case Rep 2024;29:102305) © 2024 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 circumflex aorta is an unusual vascular ring caused by a right aortic arch (RAA) with a left-sided ligamentum arteriosum and a descending aorta that crosses posteriorly from right to left above the level of the tracheal carina. The uncrossing operation offers definitive surgical correction for properly selected patients.[1]

CASE

A 9-year-old boy initially presented with persistent cough, stridor, and recurrent pneumonia secondary to a vascular ring with tracheal compression. He underwent division of the ligamentum arteriosum and posterior aortopexy. Two years later, an anterior aortopexy was performed for persistent symptoms, but to no avail. Follow-up computed tomography revealed a circumflex aorta (**Figure 1A**). Bronchoscopy showed tracheobronchomalacia and pulsatile posterior tracheal compression (Video 1). The uncrossing procedure was performed using moderate hypothermic circulatory arrest and selective antegrade cerebral perfusion. The aorta was transected distal to the head and neck vessels, and the proximal stump was oversewn. The descending aorta was dissected from its posterior attachments, "uncrossed," transposed to the left of the trachea, and anastomosed to the ascending aorta (Video 1, **Figure 1B**). Postoperative imaging revealed an intact reconstruction (**Figure 1C**) and complete resolution of tracheal compression (Video 1). The child was discharged 5 days after the operation with significant symptomatic relief.

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ABBREVIATIONS AND ACRONYMS

RAA = right aortic arch

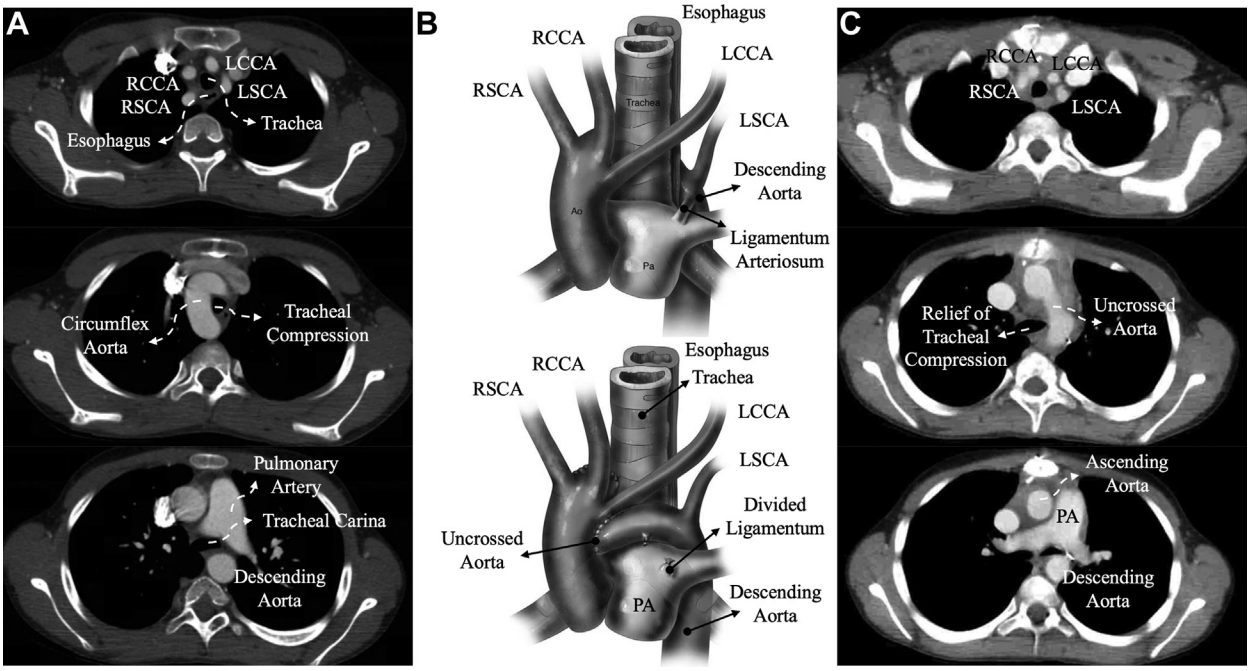
RLN = recurrent laryngeal nerve

DISCUSSION

Although circumflex aorta is uncommon, it should be included in the differential diagnosis of recurrent pediatric respiratory distress. As observed in the present case, the pertinent anatomy can sometimes be challenging to discern, leading to a missed diagnosis. For patients with clinical and imaging evidence of tracheal and esophageal compression from a circumflex aorta, the uncrossing operation offers durable relief of symptoms and is being increasingly reported with favorable outcomes.[1] Importantly, among these patients, ligamentum division alone often does not adequately relieve the posterior and right-sided tracheal compression caused by the transverse aorta and RAA, respectively.[2,3] Moreover, an unsuccessful prior vascular ring division or aortopexy may increase the complexity of the subsequent uncrossing procedure.[3]

Although the conduct of the uncrossing operation varies among institutions, several overarching technical principles deserve mention.[2] First, marking sutures placed on the left lateral side of the ascending aorta at the site of the proposed implantation before the initiation of cardiopulmonary bypass (ie, with the heart distended) may facilitate aortic uncrossing later in the operation.[1,3] Second, during dissection of the RAA to delineate the site of aortic transection distal to the right subclavian artery, caution must be exercised to avoid injury to the right recurrent laryngeal nerve (RLN). Similarly, during dissection of the descending aorta from its posterior attachments, great care must be taken in the region of the left RLN.[1,2] Finally, after aortic uncrossing, the path of the descending aorta should be inspected to ensure a tension-free anastomosis without irregularities. Furthermore, there should be sufficient room for the right pulmonary artery to traverse anterior to the

FIGURE 1 Uncrossing Operation for Circumflex Aorta



(A) Preoperative CTA reveals a circumflex aorta caused by mirror-image branching of the head and neck vessels, a right aortic arch, and the descending aorta crossing posteriorly from right to left above the carina. (B) The top schematic illustrates the anatomy of the circumflex aorta as viewed through a median sternotomy incision. The ascending aorta and aortic arch are on the right, and the descending aorta crosses posterior to the trachea and esophagus. The left subclavian artery originates from the descending aorta. The bottom schematic depicts the completed uncrossing of the aorta. The descending aorta has been brought anterior to the trachea and esophagus and anastomosed to the side of the ascending aorta, thus relieving the posterior compression of the trachea. Reprinted from *Operative Techniques in Thoracic and Cardiovascular Surgery*, 2013;18:15-31, with permission from Elsevier. (C) Postoperative CTA shows no tracheal compression, the uncrossed aorta, and the anastomosis between the descending aorta and ascending aorta. Ao = Aorta; CTA = computed tomography angiography; LCCA = left common carotid artery; LSCA = left subclavian artery; PA = pulmonary artery; RCCA = right common carotid artery; RSCA = right subclavian artery.

trachea and posterior to the ascending aorta and transverse arch.[2,3] Persistent obstructive respiratory symptoms after the uncrossing operation mandates a detailed bronchoscopic evaluation of the membranous trachea.

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REFERENCES

1. Russell HM, Rastatter JC, Backer CL. The aortic uncrossing procedure for circumflex aorta. *Oper Tech Thorac Cardiovasc Surg*. 2013;18:15–31.

2. Labuz DF, Kamran A, Jennings RW, Baird CW. Reoperation to correct unsuccessful vascular ring and vascular decompression surgery. *J Thorac Cardiovasc Surg*. 2022;164:199–207.

3. Backer CL, Monge MC, Wallen WJ, Eltayeb O. The aortic uncrossing procedure. *J Thorac Cardiovasc Surg*. 2024;167(2):413–419. https://doi.org/10.1016/j.jtcvs.2023.06.013

KEY WORDS aortic, congenital, ligamentum, stridor, surgery

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