

Surgical management of circumflex aorta associated with coarctation and tracheoesophageal fistula



Tiffany Hsiung, BS,^a Jack F. Donaghue, BA,^a Michael A. Antonchak, MD,^b Natalie Ostro, MD,^a Sujata B. Chakravarti, MD,^b Zachary Hena, MD,^b Michael J. Martinez, MD,^b Keith A. Kuenzler, MD,^c Ralph S. Mosca, MD,^a and T. K. Susheel Kumar, MD,^a New York, NY

From the ^aDepartment of Congenital Cardiothoracic Surgery, ^bDivision of Pediatric Cardiology, Department of Pediatrics, and ^cDivision of Pediatric Surgery, Hassenfeld Children's Hospital, NYU Langone Health, New York, NY.

Received for publication Oct 19, 2023; revisions received Nov 29, 2023; accepted for publication Jan 3, 2024; available ahead of print Jan 21, 2024.

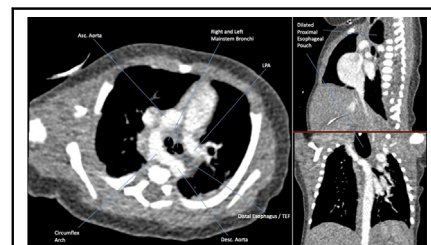
Address for reprints: T. K. Susheel Kumar, MD, Department of Congenital Cardiothoracic Surgery, NYU Langone Health, 530 First Ave, Suite 9V, New York, NY 10016 (E-mail: tkusheel.kumar@nyulangone.org).

JTCVS Techniques 2024;24:132-4

2666-2507

Copyright © 2024 The Author(s). Published by Elsevier Inc. on behalf of The American Association for Thoracic Surgery. This is an open access article under the CC BY-NC-ND license (<http://creativecommons.org/licenses/by-nc-nd/4.0/>).

<https://doi.org/10.1016/j.jtc.2024.01.009>



CT images showing the circumflex aorta and its relationship to the airway and esophagus.

CENTRAL MESSAGE

The combination of circumflex aorta, coarctation, and tracheoesophageal fistula can be a challenge for surgical correction. A staged correction may be the best approach.

▶ Video clip is available online.

Circumflex aorta (CA) is a rare form of vascular ring that can lead to significant tracheal or esophageal compression requiring surgical intervention in the form of aortic uncrossing procedure.^{1,2} To our knowledge, the association of CA with both coarctation and tracheoesophageal fistula (TEF) has not been described before. We herein describe the surgical management of right CA with coexisting coarctation and TEF in a newborn baby. Consent was obtained to publish this report; institutional review board approval was not required.

CASE

A newborn baby, born at full term weighing 3.1 kg, with prenatal diagnosis of DiGeorge syndrome, right aortic arch, and polyhydramnios was noted to have respiratory distress at birth. Postnatal course was complicated by difficulty in passing a nasogastric tube, raising suspicion for esophageal atresia. A computed tomography scan confirmed the diagnoses of right CA with an aberrant left subclavian artery and type C TEF (Figure 1). Mild-to-moderate narrowing of the trachea was also noted. Echocardiogram confirmed right aortic arch with mild coarctation of the aorta (peak gradient of 20 mm Hg) in the setting of a large ductus.

The patient underwent a flexible bronchoscopy, which demonstrated a fistulous connection of the distal trachea with the distal esophageal segment. Following a multidisciplinary discussion, the patient underwent a right

thoracotomy through the fourth intercostal space on day 2 of life. The distal esophagus was isolated and the fistulous connection to the distal trachea was divided. The proximal esophageal pouch was located behind the superior vena cava above the transverse aortic arch. Given the considerable gap between the esophageal segments and the imminent need for cardiac intervention, a decision was made to leave the upper pouch alone and perform a gastrostomy. Despite continuous evacuation of the proximal esophageal pouch, the patient could not be weaned off noninvasive mechanical ventilation due to recurrent collapse of the right lung, indicating significant airway compression from the vascular ring. In addition, a repeat echocardiogram following cessation of prostaglandin demonstrated significant narrowing of the distal arch and the isthmus (peak gradient of 79 mm Hg).

PROSTIN (Pfizer) infusion was restarted, and the decision was made to proceed to the operating room for an aortic uncrossing procedure with correction of the coarctation. The infant was placed on cardiopulmonary bypass via cannulation of the ascending aorta and right atrium

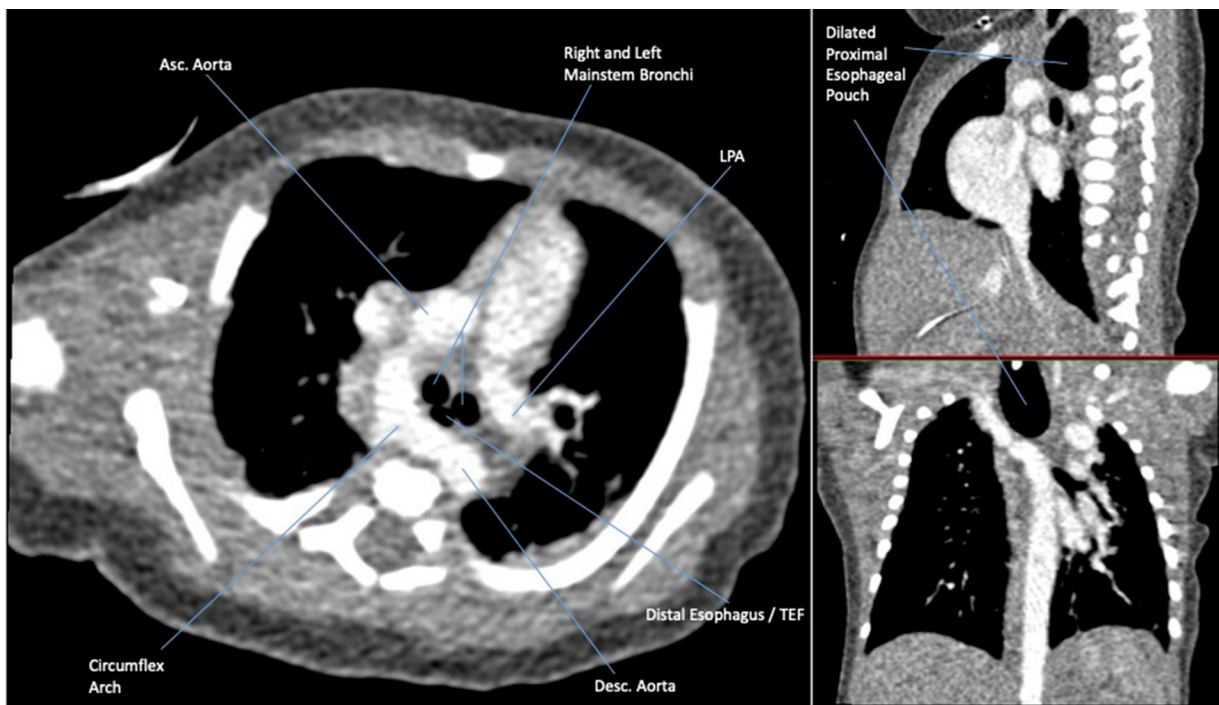


FIGURE 1. Computed tomography images showing the circumflex aorta and its relationship to the airway and esophagus.

and was cooled to deep hypothermia. The patent ductus arteriosus was transected, and the aorta was divided at the transverse arch just beyond the origin of the right subclavian artery. The proximal stump was closed, and the distal portion was anastomosed to the leftward aspect of the distal ascending aorta and the proximal left carotid artery. The aberrant left subclavian artery was sacrificed to allow mobilization of the descending

thoracic aorta. A patch of pulmonary homograft was used to augment the distal transverse arch and the proximal descending thoracic aorta, under a short period of circulatory arrest.

The patient did well in the postoperative period and was successfully extubated on postoperative day 11. Approximately 1 month later, the patient underwent repair of esophageal atresia via redo thoracotomy. The proximal esophageal pouch and the distal esophageal segments were mobilized, and esophageal anastomosis could be performed easily (Figure 2). The patient underwent balloon angioplasty for mild narrowing of the distal transverse arch at 2 months of life (Video 1). The patient was discharged home at 3 months of age on room air. At 1-year follow-up, the patient continues to do well, with good oral feeding and without any signs of esophageal or airway compression or recoarctation of the aorta.

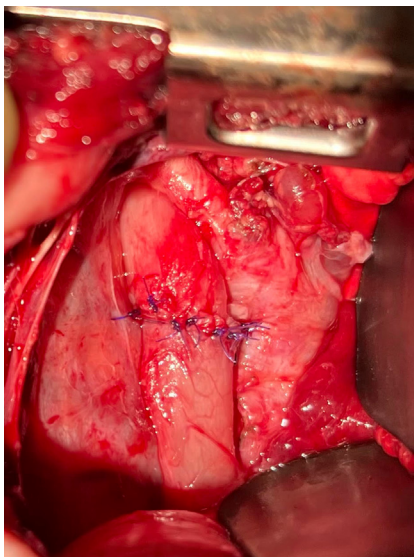
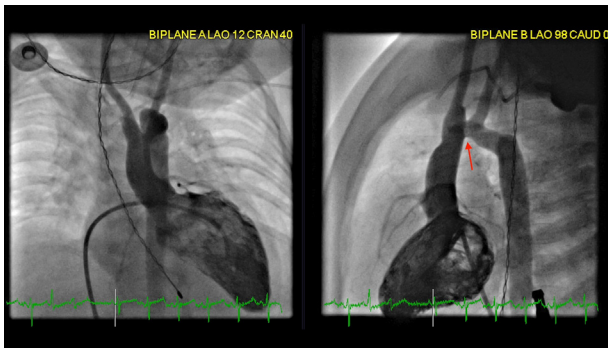


FIGURE 2. Surgical image showing completion of the staged esophageal anastomosis following the aortic uncrossing procedure.

DISCUSSION

CA is a rare vascular ring in which the transverse portion of the arch crosses the midline posterior to the esophagus, typically at or just above the level of the carina, and then descends on the contralateral side of the spine. It requires surgical intervention when there is evidence of tracheal or esophageal compression.^{1,2} Although quite rare, CA can be associated with hypoplasia of the transverse arch, as observed in our case; however, in most cases of CA, the arch is of normal caliber.^{2,3} Our patient had both hypoplasia of the



VIDEO 1. Video demonstrating surgical management of circumflex aorta associated with coarctation and tracheoesophageal fistula. Video available at: [https://www.jtcvs.org/article/S2666-2507\(24\)00011-7/fulltext](https://www.jtcvs.org/article/S2666-2507(24)00011-7/fulltext).

transverse arch (with later development of critical coarctation) and evidence of airway compression, mandating surgical intervention. Most cases of aortic uncrossing procedure have been described beyond infancy.^{1,2} The association of CA with TEF has not been described before and makes the correction more challenging. A right CA can potentially come in the way of primary esophageal anastomosis, whereas conversely it may be dangerous to proceed with aortic uncrossing procedure without correction of TEF. A staged repair as described in this report may be the best approach to correct this difficult combination of defects. In the first stage, a conservative approach that separates the trachea from the

esophagus with placement of a gastrostomy tube is adopted to make the patient a safe candidate for correction of the vascular ring. The second stage involves aortic uncrossing procedure with augmentation of the arch if needed. The third stage consists of chest reentry and esophageal anastomosis to complete correction of TEF. Although concomitant interventions on circumflex aorta and trachea have been described before, this is the first report, to our knowledge, of successful surgical management of TEF associated with CA.⁴

Conflict of Interest Statement

The authors reported no conflicts of interest.

The *Journal* policy requires editors and reviewers to disclose conflicts of interest and to decline handling or reviewing manuscripts for which they may have a conflict of interest. The editors and reviewers of this article have no conflicts of interest.

References

1. Planche C, Lacour-Gayet F. Aortic uncrossing for compressive circumflex aorta. 3 cases. *Presse Med.* 1984;13(21):1331-1332.
2. Backer CL, Mongé MC, Wallen WJ, Eltayeb O. The aortic uncrossing procedure. *J Thorac Cardiovasc Surg.* 2023;167:413-419.
3. Haranal M, Abdul-Aziz KA, Latiff HA, Srimurugan B, Sivalingam S. Aortic uncrossing with concomitant arch augmentation in a rare case of left circumflex aorta with arch hypoplasia. *Indian J Thorac Cardiovasc Surg.* 2021;37(6):673-675.
4. Nuri H, Torre M. Aortic arch uncrossing using a tracheal resection procedure. *Multimed Man Cardiothorac Surg.* 2021;2021.