Congenital absence of tracheal rings: A video case report

Clara Angeles, MD, MS,[a,b,c] Tony Kille, MD,[a,d] Joshua L. Hermsen, MD,[a,b,c] Malcolm M. DeCamp, MD,[a,b] and Petros V. Anagnostopoulos, MD, MBA,[a,b,c] Madison, Wis

From the [a]Department of Surgery, [b]Division of Cardiothoracic Surgery, [c]Section of Pediatric Cardiothoracic Surgery, and [d]Section of Pediatric Otolaryngology, Division of Otolaryngology, Head, and Neck Surgery, University of Wisconsin, Madison, Wis.

Disclosures: 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.

Read at the 103rd Annual Meeting of The American Association for Thoracic Surgery, Los Angeles, California, May 6-9, 2023.

Informed consent: The subject(s) parents provided informed written consent for the publication of the study data.

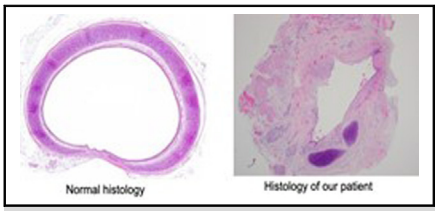
Received for publication May 5, 2023; revisions received June 23, 2023; accepted for publication June 26, 2023; available ahead of print July 22, 2023.

Address for reprints: Petros V. Anagnostopoulos, MD, MBA, Department of Surgery, The University of Wisconsin, H4/358 Clinical Sciences Center, 600 Highland Ave, Madison, WI 53792 (E-mail: petros@surgery.wisc.edu).

JTCVS Techniques 2023;21:184-7

2666-2507

https://doi.org/10.1016/j.xjtc.2023.06.019



Histology shows *left* normal C-shaped cartilaginous ring and *right* foci of cartilage in the specimen.

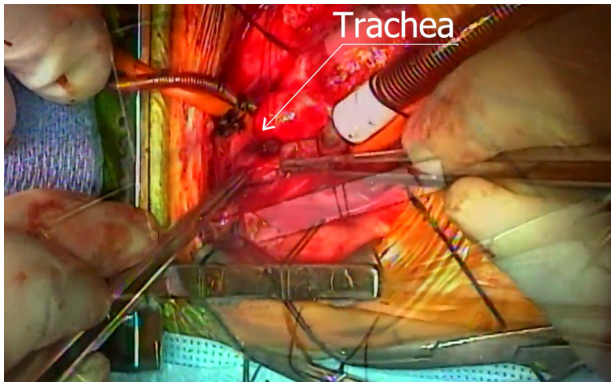
CENTRAL MESSAGE

We present a case of a neonate with functional tracheal stenosis due to congenital absence of tracheal rings who was treated successfully with a short-segment tracheal resection and anastomosis.

Video clip is available online.

Congenital tracheal stenosis (CTS) is a potentially life-threatening disorder that leads to early life severe airway compromise. Common causes include complete tracheal rings or presence of a vascular ring with secondary tracheobronchomalacia.[1] Congenital absence of tracheal rings is an exceedingly rare cause of CTS[2,3] that presents as severe focal tracheomalacia.[2] Here we present a case of a neonate with functional tracheal stenosis due to absence of tracheal rings (Video 1). The patient(s) parents provided informed written consent for the publication of the study data; institutional review board approval was not required.

A 3-week-old, ex-31-week premature female, weighing 2.5 kg, was transferred from an outside facility with tachypnea and stridor since birth. A rigid bronchoscopy had demonstrated significant narrowing of the distal trachea. A computed tomography scan revealed severe localized airway narrowing and confirmed no other cardiovascular abnormalities (Figure 1).[4] The patient was taken for bronchoscopy evaluation. She was found to have severe dynamic narrowing at the distal trachea 1 cm above the carina. The stenotic airway, measuring 5 mm in length, appeared to be compliant and distensible with positive pressure ventilation without evidence of complete tracheal rings. A 1.9-mm telescope on a 2.5-mm endotracheal tube bypassed this region. Overnight, the child decompensated requiring reintervention. The tip of the endotracheal tube had moved proximally and was no longer stenting open the narrowed



VIDEO 1. Congenital absence of trachea rings. Video available at: https://www.jtcvs.org/article/S2666-2507(23)00226-2/fulltext.

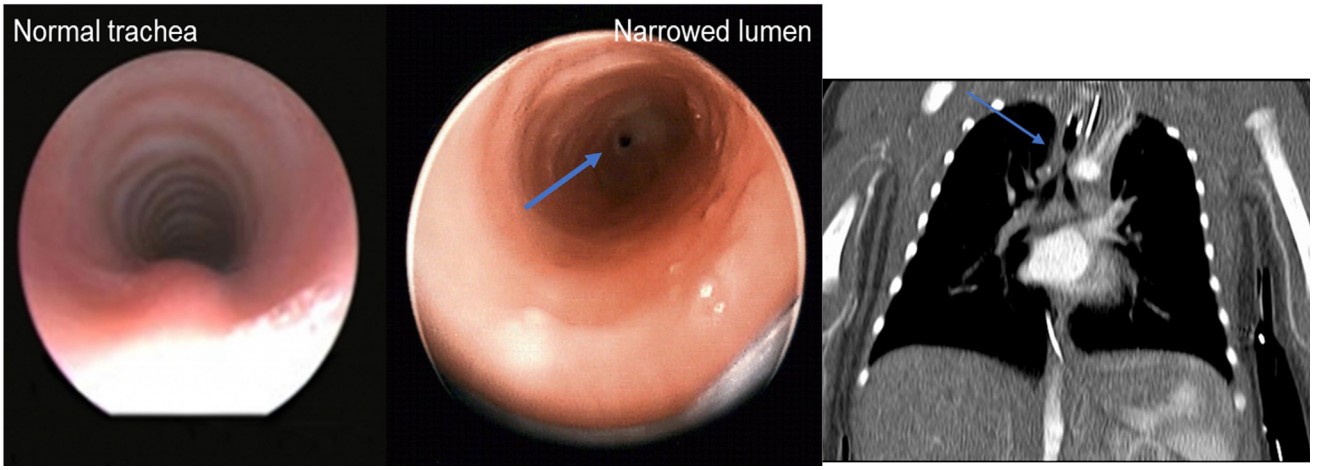


FIGURE 1. Narrowing of the distal trachea on initial bronchoscopy and computed tomography.

area. This was noted to be unsustainable and tracheal resection was planned.

After median sternotomy, cardiopulmonary bypass was initiated. Using bronchoscopy, the proximal and distal margins of the narrowing were marked. The diseased trachea was completely excised and it was noted to have absence of tracheal rings. A small incision was made anterior proximally and a mirror image incision was made posteriorly in the distal trachea at the membranous portion (Figure E1). The 2 segments were anastomosed using a posterior running 7-0 Prolene suture (Figure E2) and interrupted horizontal mattress everting anterior sutures (Figure E3). Postrepair bronchoscopy showed no evidence of residual stenosis. The patient was extubated successfully on postoperative day 1 and transferred to the neonatal intensive care unit for continued care of prematurity. Gross and histologic findings were consistent with congenital absence of tracheal rings. Figure 2 shows the comparison of a normal cartilaginous ring with the pathologic finding in the specimen of foci of cartilage embedded in respiratory epithelium with subepithelial fibrosis. The child underwent 2 subsequent bronchoscopies that showed no granulation formation or anastomotic narrowing. She was discharged home in stable conditions on postoperative day 52, equivalent to 41-week gestation. Six-month follow-up bronchoscopy, showed complete healing (Figure 3).

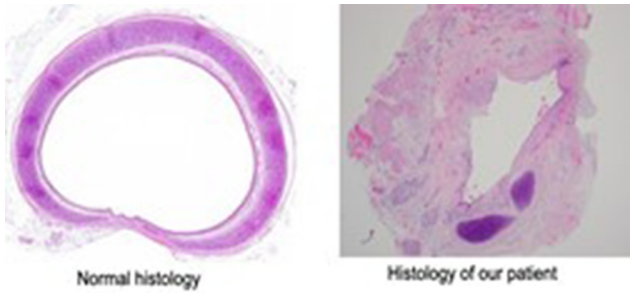


FIGURE 2. Histology shows *left* normal C-shaped cartilaginous ring and *right* foci of cartilage in the specimen.

Absent tracheal rings is an extremely rare intrinsic tracheal defect.[3] The absence of the cartilaginous rings leads to airway collapse during fluctuations of pressure associated with normal ventilation, coughing, and crying. Patients will present with stridor, difficulty breathing, and airway compromise and become symptomatic much earlier in life compared with patients with other defects. Smith and colleagues[3] presented 4 patients with congenital absence of distal tracheal rings. All were repaired successfully using a resection with slide tracheoplasty and carinal reconstruction; none required reinterventions. We elected to perform a resection with a primary spatulated anastomosis using everting sutures as described by Hobbs and colleagues.[5] The segment of abnormal trachea must be completely resected. The primary anastomosis was performed using nonabsorbable sutures (Prolene) in the membranous portion, and anteriorly interrupted absorbable horizontal mattress everting sutures (PDS; Ethicon) were used to facilitate mucosal apposition. This technique can result in an unobstructed airway and may have a lower risk of recurrence than the standard running technique. It avoids the figure-of-8 deformity and may decrease the incidence of postoperative complications and reinterventions secondary to granulation tissue formation or contraction[5] (Figure E4). Known reported complications of airway reconstruction in children include reintubation, failed extubation and tracheostomy, reinterventions for ballooning and stenting, as well as perioperative mortality.[6] This demonstrates the complexity in the treatment of these children and the need for continued improvement in surgical techniques and management of the abnormalities of the tracheobronchial tree.

This unique case highlights the importance of keeping in mind different causes of congenital tracheal stenosis in

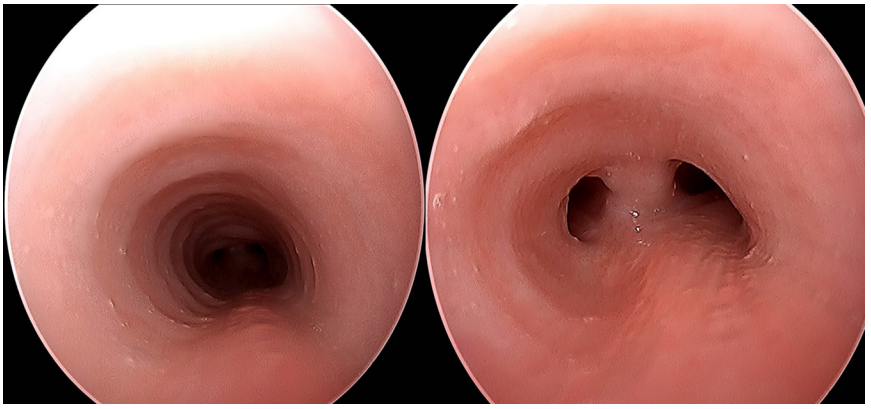


FIGURE 3. Six-month follow-up bronchoscopy.

neonates. Absence of tracheal rings is a life-threatening disorder and must be recognized as a cause of stridor and dyspnea earlier in life. Some of the critical concepts learned are that the ringless segment must be completely excised compared with other causes such as complete tracheal rings. Additionally, the use of an interrupted horizontal mattress everting suture technique can be applied during the neonatal period as well.

This case presents a premature, low-birth weight neonate who underwent a successful resection of a short segment of tracheal stenosis from absence of tracheal rings with primary anastomosis of the unaffected trachea.

References

1. Hofferberth SC, Watters K, Rahbar R, Fynn-Thompson F. Management of congenital tracheal stenosis. *Pediatrics*. 2015;136:e660-9.
2. Seymour FK, Roebuck DJ, McLaren CA, Bailey CM. Congenital segmental absence of tracheal rings. *Int J Pediatr Otorhinolaryngol*. 2006;1:45-9.
3. Smith MM, Kou YF, Schweiger C, Lehenbauer DG, de Alarcon A, Rutter MJ. Congenital absence of tracheal or bronchial rings. *Otolaryngol Head Neck Surg*. 2021;164:422-6.
4. Children's Hospital of Philadelphia, Center for Pediatric Airway Disorders. Accessed August 2, 2023. https://www.chop.edu/treatments/bronchoscopy
5. Hobbs RD, Moon J, Murala J, Ohye RG. Novel suture technique for slide tracheoplasty for the treatment of long-segment tracheal stenosis. *Semin Thorac Cardiovasc Surg*. 2020;32:930-4.
6. Stephens EH, Eltayeb O, Mongé MC, Forbess JM, Rastatter JC, Rigsby CK, et al. Pediatric tracheal surgery: a 25-year review of slide tracheoplasty and tracheal resection. *Ann Thorac Surg*. 2020;109:148-53.

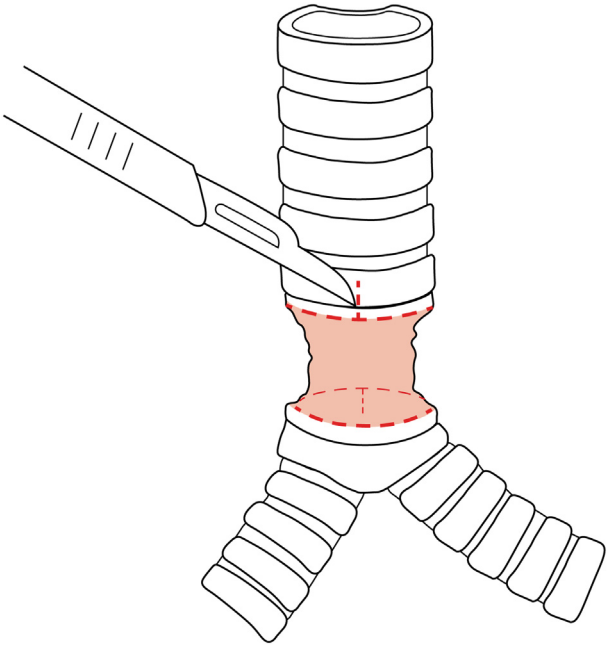


FIGURE E1. The area of diseased trachea was excised completely. An incision was made anteriorly in the proximal segment of the trachea at the 12 o'clock position and a mirror image incision was made posteriorly in the membranous trachea distally. Illustration by Margaret Greco, MD.

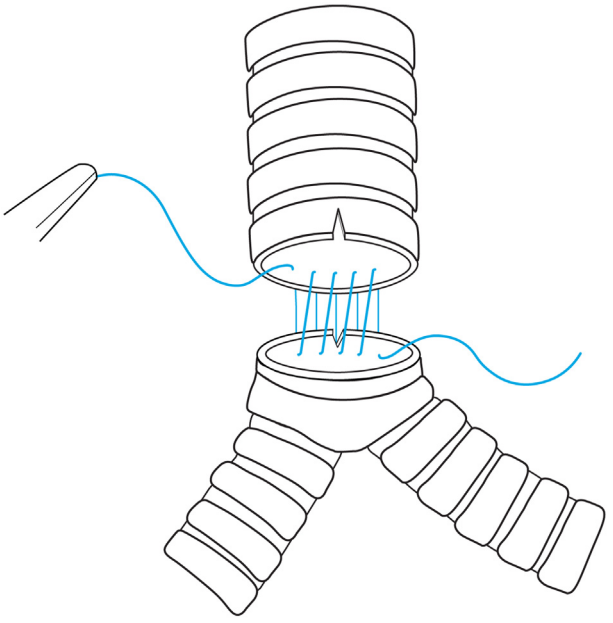


FIGURE E2. The posterior membranous tracheal elements were anastomosed using 7–0 Prolene and a running suture technique. Illustration by Margaret Greco, MD.

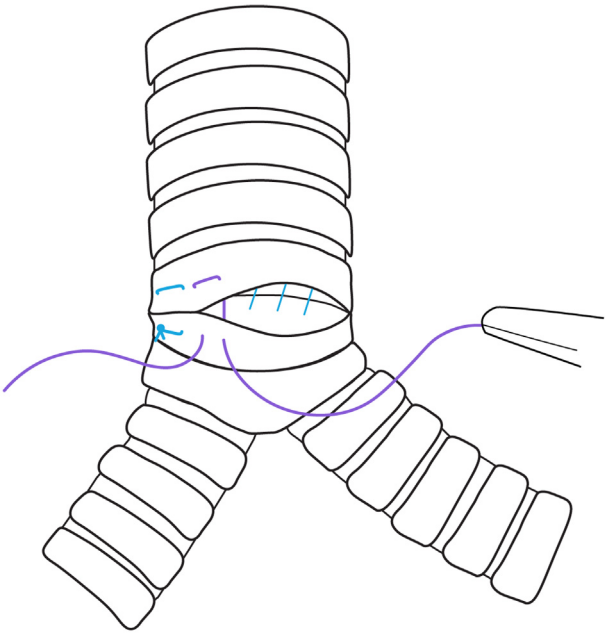


FIGURE E3. The posterior running suture was locked on either end to 7–0 Prolene everting horizontal mattress sutures. Three additional 7–0 PDS (Ethicon) everting horizontal mattress sutures completed the connection anteriorly. Illustration by Margaret Greco, MD.

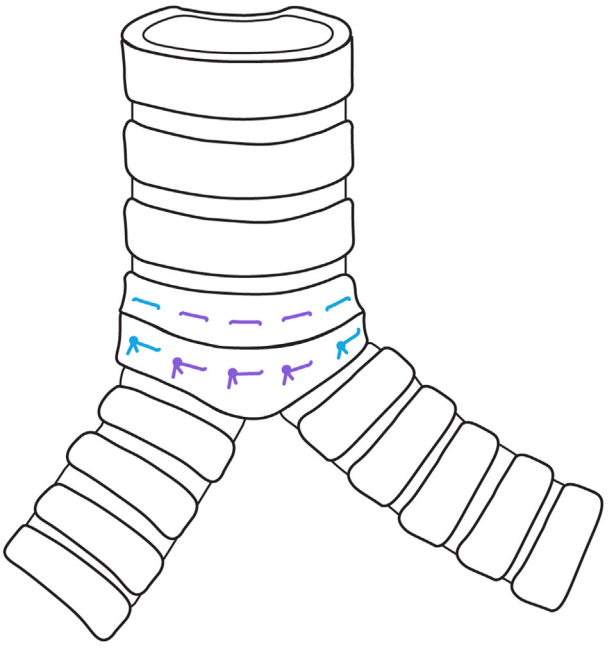


FIGURE E4. Final anastomosis. Illustration by Margaret Greco, MD.