CONGENITAL ABSENCE OF TRACHEAL RINGS – A VIDEO CASE REPORT

Clara Angeles, MD, MS 1,2,3, Tony Kille, MD 1,4, Joshua L. Hermsen, MD 1,2,3, Malcolm M. DeCamp, MD 1,2, Petros V. Anagnostopoulos, MD, MBA 1,2,3

Affiliations:

1. Department of Surgery - University of Wisconsin, Madison, Wisconsin
2. Division of Cardiothoracic Surgery - University of Wisconsin, Madison, Wisconsin
3. Division of Cardiothoracic Surgery – Section of Pediatric Cardiothoracic Surgery - University of Wisconsin, Madison, Wisconsin
4. Division of Otolaryngology- Head & Neck Surgery – Section of Pediatric Otolaryngology - University of Wisconsin, Madison, Wisconsin

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Corresponding Author:

Petros V Anagnostopoulos, MD, MBA
Surgeon-In-Chief, The American Family Children’s Hospital
Chief Pediatric Cardiothoracic Surgery,
The University of Wisconsin, Department of Surgery
H4/358 Clinical Sciences Center
Glossary Of Abbreviations

CTS: Congenital Tracheal Stenosis

ETT: Endotracheal Tube

CPB: Cardiopulmonary Bypass

POD: Postoperative Day

NICU: Neonatal Intensive Care Unit
CENTRAL PICTURE

Legend: Histology. Left normal C-shaped cartilaginous ring. Right foci of cartilage in specimen.

CENTRAL MESSAGE

We present a case of a neonate with functional tracheal stenosis due to congenital absence of tracheal rings, treated successfully with a short-segment tracheal resection and anastomosis.
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. Congenital absence of tracheal rings is an exceedingly rare cause of CTS, that presents as severe focal tracheomalacia. Here we present a case of a neonate with functional tracheal stenosis due to absence of tracheal rings (Video 1).

The subject(s) parents provided informed written consent for the publication of the study data; IRB approval was not required.

A 3-week-old, ex-31-week premature female, weighing 2.5kg, was transferred from an outside facility with tachypnea and stridor since birth. A rigid bronchoscopy had demonstrated significant narrowing of the distal trachea. A CT scan revealed severe localized airway narrowing and confirmed no other cardiovascular abnormalities (Figure 1). 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 ETT bypassed this region. Overnight, the child decompensated; requiring reintervention. The tip of the ETT had moved proximally and was no longer stenting open the narrowed area. This was noted to be unsustainable and tracheal resection was planned.
After median sternotomy, CPB 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 (Supplemental Figure 1). The two segments were anastomosed using a posterior running 7-0 Prolene suture (Supplemental Figure 2) and interrupted horizontal mattress everting anterior sutures (Supplemental Figure 3). Post repair bronchoscopy showed no evidence of residual stenosis. The patient was extubated successfully on POD1 and transferred to the NICU for continued care of prematurity. Gross and histologic findings were consistent with congenital absence of tracheal rings. The central picture 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 two subsequent bronchoscopies which showed no granulation formation or anastomotic narrowing. She was discharged home in stable conditions on POD52, equivalent to 41-week gestation. Six-month follow-up bronchoscopy, showed complete healing (Figure 2).

Absent tracheal rings is an extremely rare intrinsic tracheal defect\(^4\). 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 to patients from other causes. Smith et al\(^4\) 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, et al\textsuperscript{5}. The segment of abnormal trachea must be completely resected. The primary anastomosis was performed using non-absorbable sutures (Prolene) in the membranous portion, and anteriorly interrupted absorbable horizontal mattress everting sutures (PDS) 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-eight deformity and may decrease the incidence of post-operative complications and re-interventions secondary to granulation tissue formation or contraction\textsuperscript{5} (Supplemental Figure 4). Known reported complications of airway reconstruction in children include reintubation, failed extubation and tracheostomy, re-interventions for ballooning and stenting as well as perioperative mortality\textsuperscript{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 CTS in 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 to other causes such as complete tracheal rings. Additionally, the use of an interrupted horizontal mattress everting suture technique can be applied in 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


Figure 1: Narrowing of the distal trachea on initial bronchoscopy and CT.

Figure 2: Six month follow up bronchoscopy

Supplemental Figure 1. 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.*

Supplemental Figure 2. The posterior membranous tracheal elements were anastomosed using 7-0 Prolene and a running suture technique. *Illustration by Margaret Greco, MD.*

Supplemental Figure 3. The posterior running suture was locked on either end to 7-0 Prolene everting horizontal mattress sutures. Three additional 7-0 PDS everting horizontal mattress sutures completed the connection anteriorly. *Illustration by Margaret Greco, MD.*

Supplemental Figure 4. Final anastomosis. *Illustration by Margaret Greco, MD.*

Video 1. Congenital absence of trachea rings.