IMAGES IN PULMONARY, CRITICAL CARE, SLEEP MEDICINE AND THE SCIENCES

Congenital Vertical Tracheal Septum Misdiagnosed as Laryngomalacia

Karl Reiter¹, Johanna Kramer¹, Matthias Griese², and Birgit Kammer³

¹Pediatric Intensive Care Unit, Bronchoscopy Unit, ²Department of Pediatric Pulmonology, and ³Department of Pediatric Radiology, Hauner Children's Hospital, LMU Munich, Munich, Germany

ORCID ID: 0000-0003-1049-6655 (K.R.).

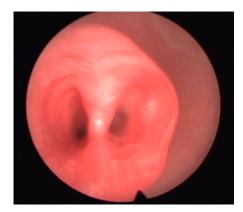


Figure 1. Bronchoscopic view on the distal trachea divided by a sagittally oriented vertical septum covered with normal mucosa. The bipartite trachea shows a smaller left- and a larger right-sided lumen. The true tracheal bifurcation is located more distally and cannot be seen in this view.

An 11-month-old girl was admitted to our hospital for the investigation of persistent stridor since birth. During her otherwise uneventful neonatal period, she had received a clinical diagnosis of laryngomalacia. On presentation, the girl was tachypneic and had jugular and intercostal retractions and a high-frequency biphasic stridor.

Bronchoscopy and chest computed tomography (Figures 1–3) revealed a distally bipartite trachea with a vertical septum almost reaching the tracheal carina, and a hypoplastic right main stem bronchus. Rigid bronchoscopy was used to allow safe mechanical ventilation during the procedure and to span the tracheal lumen to improve view after stepwise laser dissection of the septum. We chose the neodymium-doped yttrium aluminum garnet laser for deeper tissue penetration, although other laser modalities may be equally effective. Surgery was deemed to incur increased risk and was regarded as a secondary option. Surveillance bronchoscopies after 6 weeks and 3 years revealed minimal residua of the septum, tracheomalacia, and malformed tracheal cartilages confined to the distal trachea (Figure 4). Clinical follow-up showed absent dyspnea and satisfactory thriving, but frequent respiratory infections resulting from impeded airway clearance. Further interventional treatments were not necessary.

Tracheal developmental disturbances include laryngotracheal clefts, tracheomalacia, tracheal stenosis/agenesis, and tracheoesophageal fistulae (1, 2). A distal bipartite trachea has not been reported before. ■







Figure 2. Chest computed tomography: axial (A), frontal (B), and sagittal (C) corresponding images reveal a vertically and sagittally oriented septum in the distal trachea (arrows). The length of the septum is underestimated by computed tomography compared with the tracheoscopic findings because no computed tomography plane fully aligns with the plane of the septum.

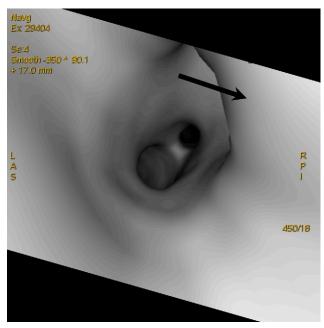


Figure 3. Chest computed tomography three-dimensional reconstruction showing the tracheal septum on the right (arrow) ending proximal to the tracheal carina, which can be seen distally. The ostium of the right main stem bronchus is hypoplastic.

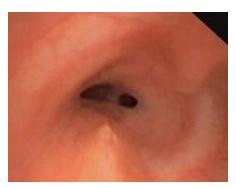


Figure 4. A surveillance bronchoscopy performed 3 years after laser treatment shows minor residua of the septum, moderate tracheomalacia, abnormal tracheal cartilage shape, and a hypoplastic right main stem bronchus.

<u>Author disclosures</u> are available with the text of this article at www.atsjournals.org.

References

- Landing BH, Dixon LG. Congenital malformations and genetic disorders of the respiratory tract (larynx, trachea, bronchi, and lungs). Am Rev Respir Dis 1979;120:151–185.
- Varela P, Torre M, Schweiger C, Nakamura H. Congenital tracheal malformations. *Pediatr Surg Int* 2018;34:701–713.