Laryngotracheal cleft in an infant with complex congenital cardiac defects

Katelin Stamm, DO, Phil Muellerleile, MD, Naila Ahmad, MD, Marion Svendrowski, MD

Department of Anesthesiology and Critical Care Medicine, Saint Louis University School of Medicine, St. Louis, Missouri

Introduction

Laryngotracheal clefts (LC), a congenital deformity of the posterior portion of the larynx and trachea, are a rare and often challenging congenital airway malformation. Occurring in every 10,000-20,000 live births, LCs cannot be diagnosed prenatally, and are usually found when symptoms manifest. We present a multi-staged case study of a neonate requiring surgical correction of cardiac anomalies with a laryngeal cleft that was diagnosed on initial intubation attempts.

Case Report

Our patient is a one day-old full term female with total anomalous pulmonary venous return diagnosed prenatally. She presented for repair of TAPVR on day one of life. She was induced and ventilated uneventfully. A 3.0 mm cuffed endotracheal tube was introduced into the trachea and appeared to be main-stemmed.

Case Report Continued

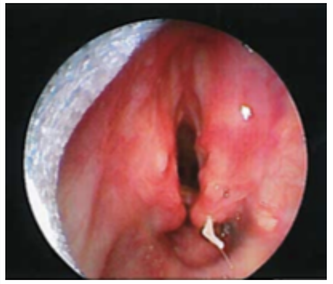
When the ETT was withdrawn a large leak was noted. Further attempts produced similar results. A 3.0 mm ETT was then main-stemmed in order to ventilate the patient. An emergent bronchoscopy revealed a Type III laryngeal cleft. Due to the urgent need for cardiac repair and difficulty in ventilating, the ENT surgeon performed a repair of the LC. Due to the urgent need for cardiac repair and difficulty in ventilating, the ENT performed a repair of the LC. The patient was transported to the ICU with a 3.0 mm uncuffed ETT in place. On day 9 of life, she underwent cardiac repair. Post-operatively, she was difficult to ventilate and her chest x-ray showed white-out of her left lung. Repeat bronchoscopy identified that her cleft repair had broken down due to prolonged intubation.

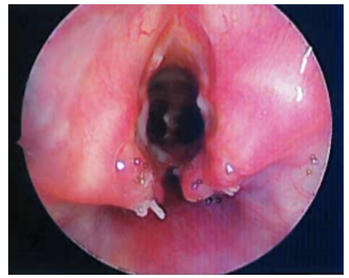
She was placed on ECMO support due to ventilatory and hemodynamic instability. There was continued difficulty with endotracheal tube placement due to a deep cleft and short trachea so she returned to the OR for a bronchoscopy, while still on ECMO.

Under direct visualization a 2.5 mm uncuffed ETT was placed in the right mainstem and another was placed in the left mainstem. A Y-adaptor was connected to both tubes and both lungs were ventilated.

Discussion

Laryngeal clefts have been characterized by a modified Benjamin and Ingles scale into five types (O-IV) ranging from a submucosal cleft (O) to a cleft which may extend to the carina (IV). With type III, or IV LCs, surgical repair is often complicated, requiring multiple revisions, with a high rate of morbidity and mortality. Proper ventilation is of utmost importance in these patients, to prevent aspiration and pulmonary complications.





If significant respiratory distress necessitates endotracheal intubation, then great care should be taken when placing the ETT, as proper placement can be difficult. Bronchoscopic guidance should be used to guide positioning. Early extubation is encouraged in repaired LCs because prolonged intubation can delay healing of the repair site.

Conclusion

Laryngeal clefts are a challenging congenital airway deformity Difficulties in endotracheal tube placement and ventilation frequently occur in more significant clefts. Creative solutions such as ETTs in each mainstream bronchus may be necessary to achieve adequate ventilation and oxygenation.

References

Leboulanger, N. et al. Laryngo-tracheo-oesophageal clefts. Orphanet Journal of Rare Diseases. 2011; 6: 81.

Mok, Quen. Airway Problems in Neonates—A Review of the Current Investigation and Management Strategies. Frontiers in Pediatrics. 2017; 5:60.

Sandu K, Monnier P. Endoscopic laryngotracheal cleft repair without tracheotomy or intubation. Laryngoscope. 2006; 116: 630-634.

Sonmez, K. et al. Our experience in two cases of type IV laryngotracheoesophageal cleft (LTEC) with a diagnosis of antenatal esophageal atresia. The Pan African Medical Journal, 2017; 26: 55.