Adult Recurrence of a Type IIIb Laryngotracheoesophageal Cleft (LTEC)

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Introduction

- Laryngotracheoesophageal clefts (LTEC) are rare congenital malformations, affecting 1 in 10,000 to 1 in 20,000 live births.[1]
- LTECs result from incomplete development of the posterior laryngeal structures and the tracheoesophageal septum.[2]
- Patients present with respiratory and gastrointestinal issues, including stridor, cyanosis, aspiration, and regurgitation.[2,3]
- The Benjamin-Inglis system classifies clefts into four types, with treatment strategies varying from endoscopic to open surgery depending on the severity.[2]
- This case report focuses on a rare instance of a Type IIIb laryngeal cleft, characterized by a complete cricoid defect extending into the extrathoracic trachea.

History and Initial Evaluation

- A 22-year-old male (born at 36 weeks) with a reported history of tracheoesophageal fistula repaired in infancy. Required feeding tube until age 9 months.
- Reported worsening dysphagia previous year, with difficulty swallowing solids more than liquids
- 5–7 pounds weight loss
- Prior evaluation by Gastroenterology (GI)
  - EGD: reflux esophagitis, hiatal hernia and Schatzki's ring
  - No improvement with dilation or PPI
  - Esophagram: significant reflux and aspiration at level of piriform sinuses
  - Gastric Emptying Study: delayed gastric emptying at 2-4 hours
- Esophagram: Esophageal reflux and aspiration from the piriform sinuses.
- Unremarkable head and neck exam and laryngoscopy. He bore none of the surgical stigmata of tracheoesophageal fistula repair, raising suspicion that the original defect was a LTEC and not a TEF.
- EAT-10 = 36

Additional Workup

- Repeat EGD with HRM: immotile esophagus (Figure 2)
- CT imaging: dehiscence of the posterior cricoid plate and laryngeal musculature (Figure 3)
- Direct microlaryngosopy (DML): Type IIIb LTEC with approximately 8 mm extension into the proximal esophagus. Significant bilious secretions were noted in the esophagus (Figure 1)

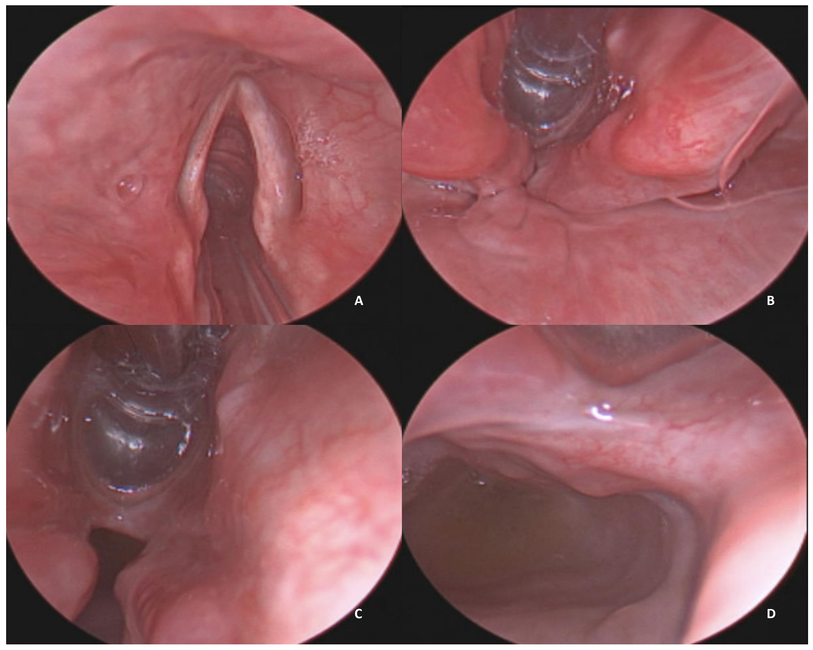

Figure 1. (A) Interarytenoid gap. (B) Dehiscence extending into cricoid. (C) Extension of LTEC into proximal trachea/esophagus. (D) Dilated esophagus with bilious reflux

Operative Management

- A gastrojejunostomy was placed pre-operatively to maximize nutrition and reduce reflux peri-operatively
- Tracheostomy at the time of surgery for airway management
- A three-layered repair was performed via an anterior laryngotracheal fissure
  - Tracheal esophageal mucosal flaps elevated
  - A fascia lata graft (5 × 7 cm) interposed between the intraluminal and extraluminal mucosal flaps
- J-tube feeding initiated on postoperative day (POD) 2.
- Return to OR POD 41 for DML
  - Well healed laryngofissure (Figure 4)
  - Residual Type I cleft
  - Decannulated
- MBSS showed moderate pharyngeal dysphagia. Therapy initiated
- Resumed total oral diet by 5 months post-op

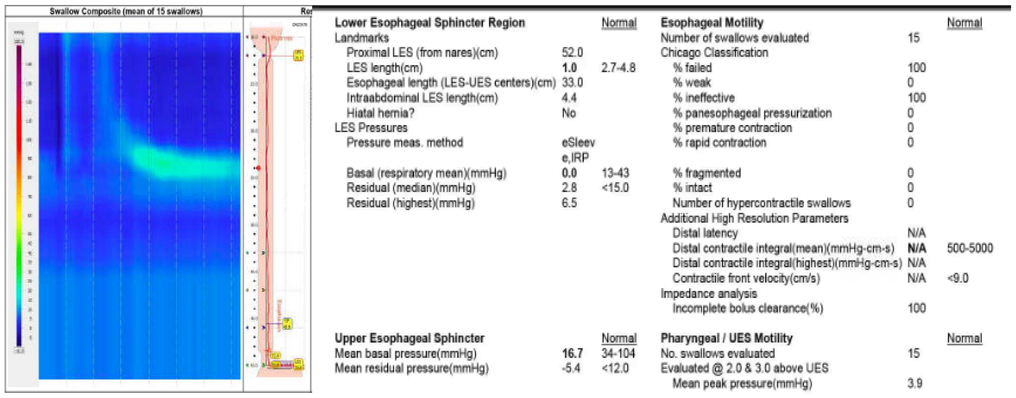

Figure 2. HRM at time of EGD. Patient has an immotile esophagus with 100% failed swallows

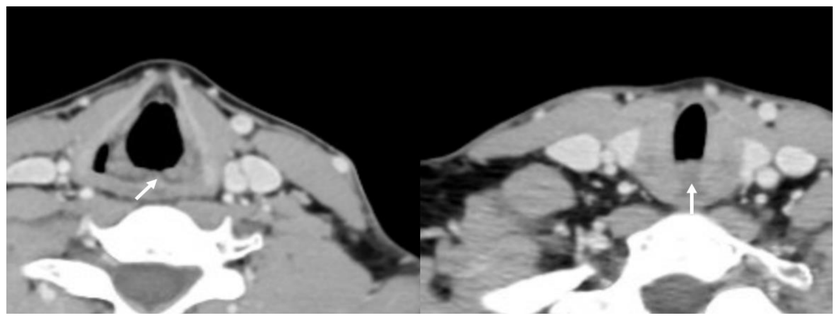

Figure 3. CT imaging demonstrating dehiscence of the musculature of the posterior larynx and tracheoesophageal party wall (white arrows).

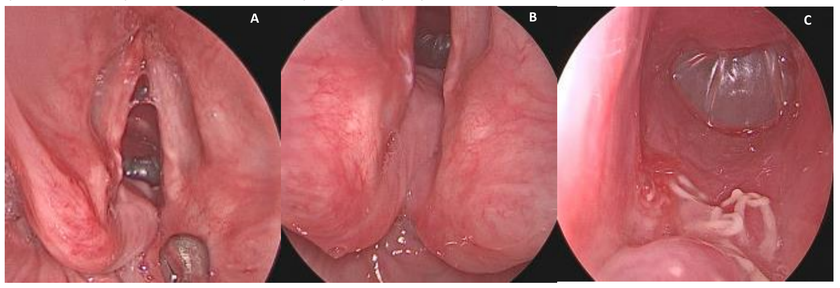

Figure 4. (A) Post-operative DML showing healed laryngofissure, **(B)** residual Type I cleft and **(C)** intact posterior tracheal wall.

Key Takeaways

- Despite advancements in in endoscopic approaches for type III LTEC, long-term outcomes for surgical repair and swallow outcomes remain uncertain
- This is the first reported case of documented recurrence in adulthood
- The patient presented with worsening dysphagia and evidence of global foregut dysfunction [reflux disease (GERD), gastroparesis and an immotile esophagus., suggesting that unresolved foregut abnormalities might contribute to surgical failure, as well as ongoing dysphagia issues.[4,5]
- Tensor fascia lata was used due to its strength, pliability, and resistance to breakdown, crucial for minimizing dehiscence in complex repairs.
- This case highlights the importance of long-term follow-up for patients with repaired laryngeal clefts, particularly those with a history of severe defects and congenital comorbidities.

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