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ORIGINAL ARTICLE

Repair of type IV laryngotracheoesophageal cleft (LTEC) on ECMO

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Abstract

Purpose A type IV laryngotracheoesophageal cleft (LTEC) is a very rare congenital malformation. Type IV LTEC that extends to the carina have poor prognosis and are difficult to manage. We present our experience with surgical repair in such a case using extracorporeal membranous oxygenation (ECMO).

Methods A male infant, who was diagnosed with Goldenhar syndrome, showed severe dyspnea and dysphagia. Laryngoscopy indicated the presence of LTEC. The patient was transferred to our institute for radical operation 26 days after birth. Prior to surgery, a balloon catheter was inserted in the cardiac region of stomach through the lower esophagus to block air leakage, to maintain positive pressure ventilation. We also performed observations with a rigid bronchoscope to assess extent of the cleft, and diagnosed the patient with type IV LTEC. After bronchoscopy, we could intubate the tracheal tube just above the carina. Under ECMO, repair of the cleft was performed by an anterior approach via median sternotomy.

Results The patient was intubated via nasotracheal tube and paralysis was maintained for 2 weeks, using a muscle relaxant for the first 3 days. Two weeks after surgery, rigid bronchoscopy showed that the repair had been completed, and the tracheal tube was successfully extubated without tracheotomy.

Conclusions Although insertion of a balloon catheter is a very simple method, it can separate the respiratory and digestive tracts. This method allowed for positive pressure ventilation and prevented displacement of the endotracheal tube until ECMO was established. As a result, we safely performed the operation and the post-operative course was excellent.

Keywords Laryngotracheoesophageal cleft · Dyspnea · Dysphagia · Rigid bronchoscopy · Extracorporeal membranous oxygenation

Introduction

Laryngotracheoesophageal cleft (LTEC) is a very rare congenital malformation characterized by a posterior midline deficiency where the larynx and trachea separate to form the hypopharynx and esophagus [1]. The most commonly accepted classification system was described by Benjamin and Inglis, where type I clefts are interarytenoid, at or below the level of the vocal process; type II exhibits a partial extension through the posterior cricoid plate; type III extends completely through the cricoid plate; and type IV extends through the larynx, into the cervical or intrathoracic trachea and esophagus [2]. The severity of LTEC is directly correlated with the downward extension of the cleft severity; symptoms range from mild stridor to massive aspiration and respiratory distress [3, 4]. The reported incidence varies from 1 in 10,000 to 1 in 20,000 live births, with greater frequency in males. However, it is difficult to estimate the actual birth frequency because low-grade LTEC may either be asymptomatic or show only mild symptoms; conversely, high-grade LTEC has a high mortality rate, often leading to the patient's death before diagnosis can be made [4]. In fact, a recent study, conducted between 2002 and 2005, found 20 cases of type I LTEC (7.6%) among 264 patients with chronic cough and/or aspiration [5].

If a patient is suspected of LTEC, the diagnosis is confirmed by rigid bronchoscopy. Identifying the distal extent of the cleft is essential to support the child [6]. Most type I clefts do not require surgical intervention and can be managed conservatively [5, 7]. When surgical intervention is

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required for small clefts, endoscopic repair is successful in over 80% of cases [8, 9]. In addition, endoscopic surgery for LTEC has been applied to type III LTEC [8, 10, 11]. However, in a type IV LTEC, maintenance of ventilation during bronchoscopy and subsequent surgical repair can be difficult [6]; any attempt to reconstruct type IV LTEC also involves high risks of mortality and morbidity, as well as a long hospital stay [12]. Ryan and Doody suggested that if the cleft extends into the carina or beyond, maintenance of the airway with a large cuffed endotracheal tube placed in the common tracheo-esophagus, along with positive pressure ventilation, may be the sole method to stabilize the child for operative repair. However, positive pressure ventilation via this method can result in gaseous distension of the stomach [6].

The purpose of this report was to describe the ideal way of maintaining positive pressure ventilation of type IV LTEC during the perioperative period.

Patients and methods

Institutional Review Board approval (no. R30-26) was obtained for this case report.

A male neonate (birth weight 3177 g, born at 38 weeks' gestation) with a prenatal diagnosis of esophageal atresia showed excessive amniotic fluid and poor confirmation of gastric bubble. After birth, the gastric tube could be inserted without resistance and esophageal atresia was excluded. The patient was diagnosed with Goldenhar syndrome based on unilateral head and neck malformation; he was placed under spontaneous respiration on a synchronized inspiratory positive airway pressure (SiPAP), and a gastric tube was inserted due to dysphagia. However, laryngoscopy revealed that the gastric tube returned to the gastrointestinal tract after straying into the trachea. Computed tomography revealed a tracheoesophageal fistula in the upper trachea; therefore, LTEC was strongly suspected (Fig. 1). He was then transferred to our institution for surgical treatment at 26 days after birth.

Operative procedure

Prior to surgery, an 8-Fr balloon catheter was inserted in the cardiac region of stomach through the lower esophagus to block air leakage and maintain positive pressure ventilation. Rigid bronchoscopy was performed to assess the cleft extent under general anesthesia without spontaneous respiration, and the patient was diagnosed with type IV LTEC (Fig. 2).

After bronchoscopy, the tracheal tube could be intubated immediately above the carina for stenting of the full length of the cleft. Under extracorporeal membranous oxygenation (ECMO), repair of the cleft was performed by anterior approach via median sternotomy. A midline incision of the trachea was made from the thyroid cartilage (just below the anterior commissure) to the carina. The balloon catheter could be identified via LTEC, 3 cm in length. After incision and division of the marginal mucosa of the cleft, we facilitated a two-layer closure. The anterior wall of the esophagus and posterior wall of the trachea were closed with 5-0 vicryl interrupted sutures, placing knots in the lumen. The superior limit of the cleft closure was approximately at the level of the vocal cord of the arytenoids (above the transverse and oblique arytenoid muscle). Finally, the anterior tracheal wall was closed with interrupted 5-0 vicryl sutures, with nasotracheal tube intubation to secure the endotracheal lumen.

Results

To avoid straining of the anastomotic site, the patient was intubated with a nasotracheal tube and paralysis was maintained for 2 weeks after surgery, using a muscle relaxant for the first 3 days after surgery. Two weeks after surgery, rigid bronchoscopy showed that the repair had been completed, and the nasotracheal tube was successfully extubated (Fig. 3). Oxygen administration became unnecessary on day 6 after extubation. Furthermore, wheeze and stridor were not observed.

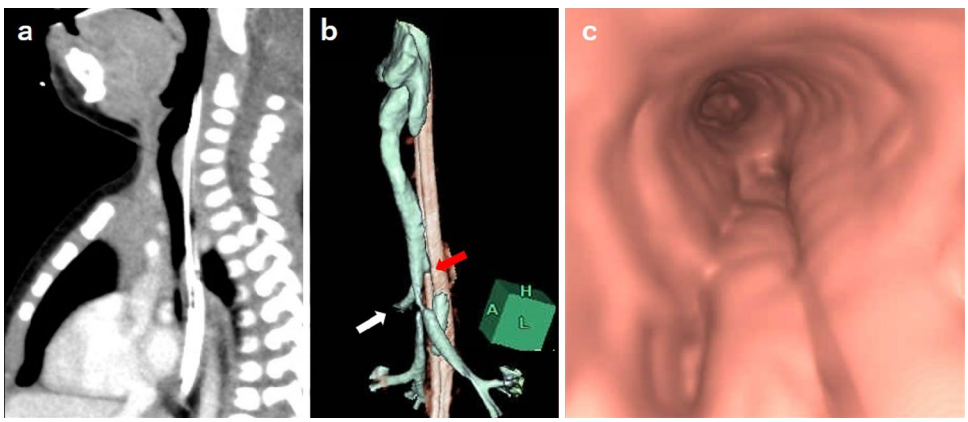

Fig. 1 Pre-operative CT findings. **a** Sagittal section. **b** LTEC (red arrow) reached just above the carina. Tracheal bronchus (white arrow). **c** Virtual bronchoscopic image shows LTEC

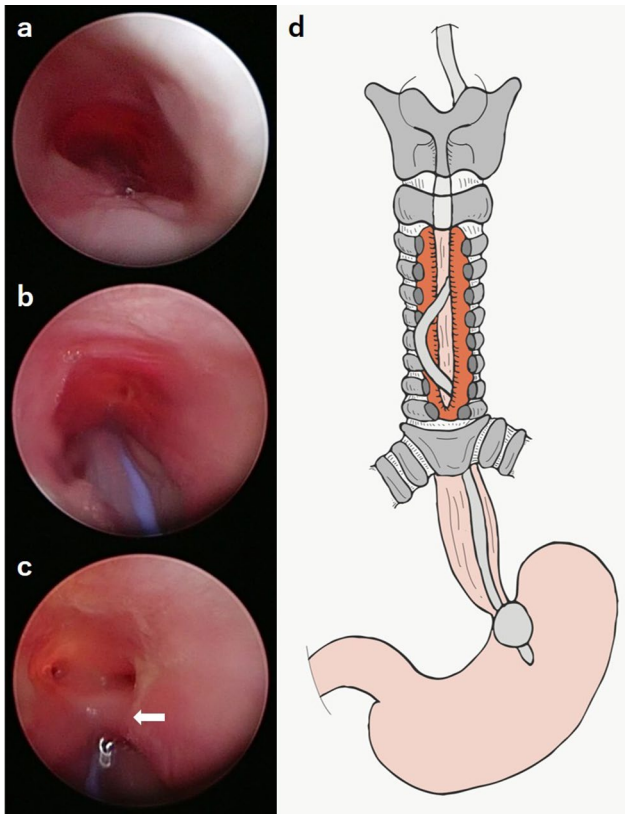

Fig. 2 Rigid bronchoscopy findings and schema of the laryngotracheal region. **a** At the level of the subglottic area, the mucosal limbs on both sides overlapped and covered the cleft. **b** The total length of the LTEC was 3 cm, and a balloon catheter inserted into the esophagus was observed through the cleft. **c** The lower edge of the LTEC (white arrow). **d** Schema of the laryngotracheal region. A balloon catheter provided ideal management to separate the respiratory and digestive tracts, thus maintaining positive pressure ventilation

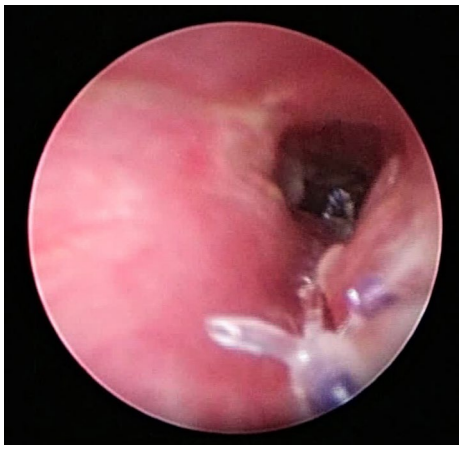

Fig. 3 Rigid bronchoscopy findings, 2 weeks after repair of the LTEC. Airway patency was maintained

On the 21st post-operative day, a remarkable gastroesophageal reflux was observed in the upper gastrointestinal series, but there was no recurrence of tracheoesophageal fistula. Laparoscopic fundoplication was performed on the 28th post-operative day. A tracheal tube of appropriate size could be inserted and there were no complications related to general anesthesia during surgery. Video fluoroscopic swallowing study showed that the patient could drink thickened liquid without any trouble. However, when he drank thin liquid, he experienced frequent aspiration and his cough reflex was absent. Although his respiratory condition improved after repair of the LTEC, he was fed by gastric tube because the swallowing dysfunction remained.

Discussion

Laryngotracheoesophageal cleft is a rare congenital anomaly that is difficult to treat. There may be a correlation between the length of the cleft and its severity and prognosis. The primary goal of treatment is to minimize respiratory complications related to aspiration, as well as to resolve feeding difficulties [13]. In this case, contrary to length of cleft, the pre-operative respiratory condition was mild; this might be because of the overlap of mucosal limbs covering the LTEC as in normal anatomy. This structure could have enabled breath management by SiPAP and tube feeding.

In a type IV LTEC, fixing an endotracheal tube is difficult. Tagawa et al. reported the successful anesthetic management of a neonate with type IV LTEC using this method [14]. Prior to surgery, we inserted a balloon catheter in the cardiac region of stomach through the lower esophagus to block air leakage; this could maintain positive pressure ventilation. Although gas from positive pressure ventilation flowed into the stomach, it was possible to reduce distension of the stomach via use of the catheter. Furthermore, the balloon catheter prevented the endotracheal tube from slipping over the upper edge of the tracheoesophageal septum into the esophagus. This method is safe, easy, and effective for separating the respiratory and digestive tracts, similar to the method of TEF block using a Fogarty catheter among patients with type C esophageal atresia; moreover, it provided maintenance of positive pressure ventilation until ECMO was established.

Because of the LTEC extended to the carina, we chose an anterior approach under ECMO. The anterior approach provided excellent visualization of the larynx for reconstruction in that area and prevented damage of recurrent and superior laryngeal nerves. Mathur et al. reported that it is preferable to manage the airway with a nasotracheal or orotracheal tube during the post-operative period, with the child maintained in a state of paralysis [12]. As a result of intubation for 2 weeks, without using interposition graft, there was no

recurrence of LTEC and tracheomalacia, and tracheotomy was not necessary.

The presence of continued swallowing dysfunction may be interpreted as unsuccessful repair of the LTEC. Osborn et al. stated that swallowing dysfunction after repair is multifactorial and arises from concomitant neurologic, anatomic, or other comorbidities that may contribute to oropharyngeal and pharyngeal dysphagia [15]. Strychowsky et al. also showed that swallowing dysfunction among patients with laryngeal cleft was caused by dysfunction at all stages of swallowing (oral phase and triggering of the swallow reflex, as well as the pharyngeal and esophageal phases), rather than simply aspiration [16]. In addition, long-term recovery of swallowing function after surgery is likely to be due to multiple factors, including surgical intervention correcting the anatomic abnormality, as well as interdependence of neurodevelopmental, anatomic, and medical factors [17]. Swallowing function is acquired with integration of various and repeated sensor-motor experience. Therefore, it is desirable to begin dysphagia rehabilitation as early as possible in patients who exhibited improved respiratory function after repair of the LTEC.

Compliance with ethical standards

Conflict of interest The authors declare that they have no conflict of interest.

Ethical approval This article does not contain any studies with human participants performed by any of the authors. This article does not contain any studies with animals performed by any of the authors.

Informed consent Informed consent for publication was obtained from the guardians of the patient in this case report, as potentially identifying information may be included in this article.

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