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Laryngotracheal separation for a type 4 laryngotracheoesophageal cleft with multiple significant malformations



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ABSTRACT

Laryngotracheoesophageal cleft (LTEC) is an extremely rare congenital malformation that is difficult to treat. We present a female patient with type 4 LTEC associated with multiple significant malformations. She suffered from a severe asthma attack due to aspiration of saliva, and it was determined that she should undergo surgery for associated congenital heart disease as early as possible to ensure long-term survival. Therefore, we performed laryngotracheal separation with an end-tracheostomy to completely and immediately prevent aspiration. She was then able to undergo radical surgery for her congenital heart disease. She recovered well postoperatively, and long-term survival is expected.

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Laryngotracheoesophageal cleft (LTEC) is an extremely rare congenital malformation in which a cleft exists in the tracheoesophageal septum. Although several cleft-closing techniques have been reported, the prognosis can be very poor depending on the severity of the LTEC and the associated malformations [1]. In particular, there are few reports of long-term survival among patients with type 4 LTEC with type C esophageal atresia. We report the successful treatment of a case of type 4 LTEC associated with multiple severe malformations. To the best of our knowledge, this is the first report of type 4 LTEC associated with type C esophageal atresia, duodenal atresia, and a double outlet right ventricle (DORV). Many classifications of LTEC have been proposed; we use the Benjamin and Inglis classification [2] in this report.

1. Case report

A female neonate was delivered via cesarean section at a gestational period of 34 weeks as a second child of a twin birth. She

had a diagnosis of suspected congenital heart disease and duodenal atresia before birth. Her birth weight was 1704 g, and her Apgar score was 0 and 9 at 1 and 5 min, respectively. Her sibling had a birth weight of 1800 g and no apparent abnormalities. After birth, the prenatally suspected duodenal atresia and congenital heart disease were confirmed, the latter of which was diagnosed as a DORV. Soon after birth, she developed respiratory distress and required intubation. Insertion of a nasogastric tube was attempted, but it strayed into the left bronchus (Fig. 1a). Contrast medium was injected into the tube in the upper esophagus; the upper esophagus, trachea, bilateral bronchi, and a tracheoesophageal fistula (TEF) was simultaneously delineated (Fig. 1b and c). Therefore, type D esophageal atresia was suspected.

First, she underwent repair of the duodenal atresia and establishment of a gastrostomy at the age of 0 days. During this surgical procedure, ventilation under general anesthesia was difficult because of a large air leak via the TEF. Banding of the intra-abdominal esophagus was not performed during this operation. She was fed via the gastrostomy, and normal growth was expected; however, severe pneumonia developed at 3 months of age because of reflux from the stomach via the TEF. To prevent this reflux, a tracheoesophageal fistulectomy was performed at 4 months of age. Positive-pressure ventilation was made possible using a Fogarty catheter to obstruct the TEF. Reflux-induced pneumonia did not recur after this procedure.

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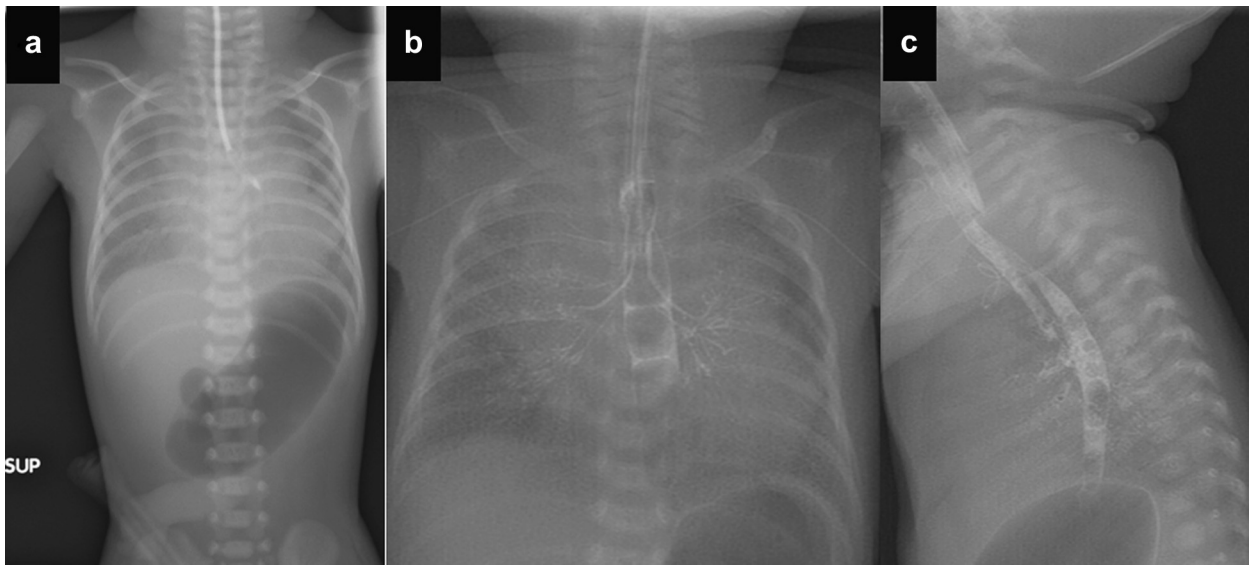


Fig. 1. Simple X-ray image and upper gastrointestinal series immediately after birth. (a) Simple X-ray. Duodenal atresia was suspected, and the nasogastric tube strayed into the left bronchus as insertion was attempted. (b) Upper gastrointestinal series, anterior position. (c) Lateral position. The esophagus, trachea, and bilateral bronchi were delineated at the same time.

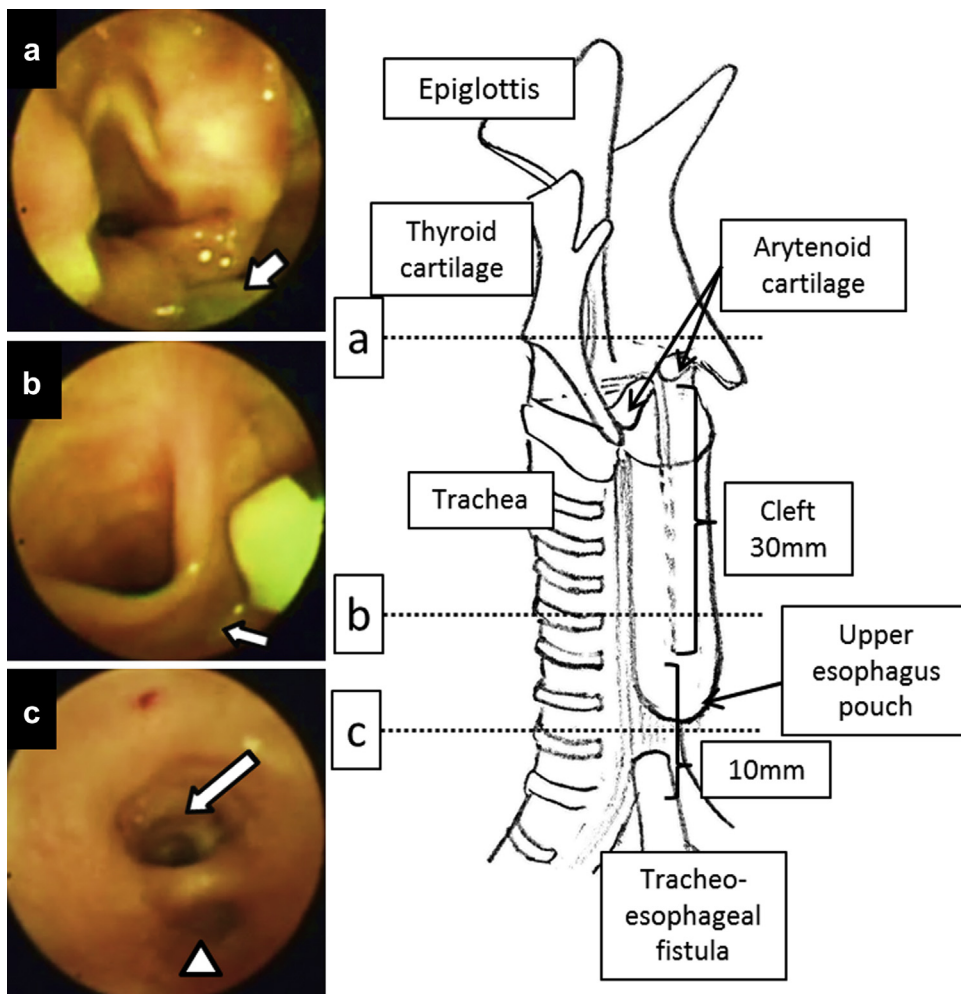


Fig. 2. Bronchoscopy findings and schema of the laryngotracheal region at 6 months of age. (a) Findings in the larynx. The endotracheal tube (arrow) is in the esophagus via the cleft. (b) The lower edge of the esophageal cleft, as viewed from the esophageal side. The area marked by the arrow is the blind end of the upper esophageal pouch. (c) The peripheral trachea, near the carina. The arrow shows the main carina, while the arrowhead shows the tracheoesophageal fistula (transsected in a previous surgery).

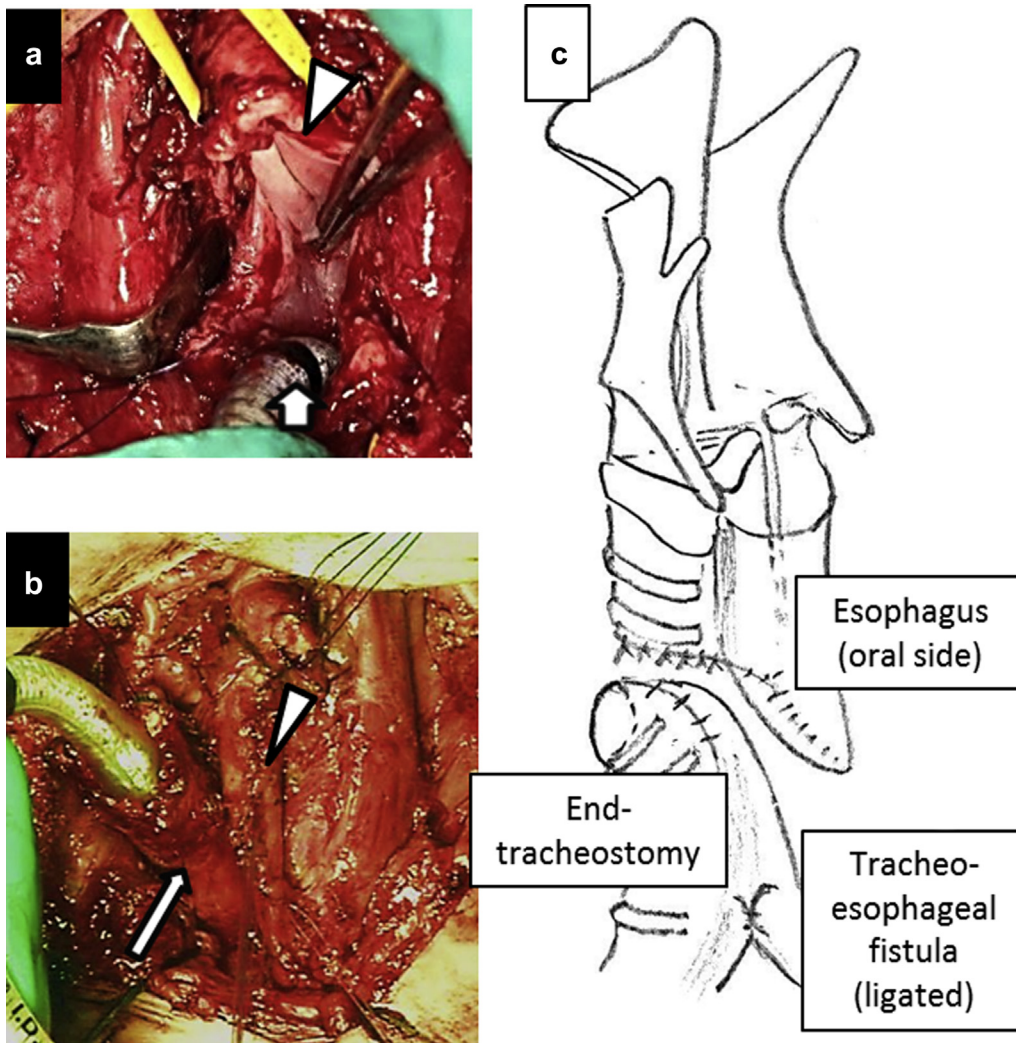


Fig. 3. Laryngotracheal separation surgery. (a) The trachea was transected at the tracheostomy. The arrowhead shows the oral side of the tracheal stump, while the arrow shows the intubation tube inserted into the peripheral side of the trachea. The forceps are inserted into the upper esophageal pouch. (b) Trachea and esophagus after reconstruction (the arrow shows the trachea, and the arrowhead shows the esophagus). (c) Schema of the completed laryngotracheal separation. The larynx was closed, and an end-tracheostomy was established.

At the age of 6 months, bronchoscopy under general anesthesia clearly showed an LTEC extending into the thoracic trachea 1 cm above the main carina, and type C esophageal atresia was present (Fig. 2) rather than type D esophageal atresia. At the age of 11 months, a tracheostomy was established to remove secretions from the airway; however, aspiration of saliva into the respiratory tract resulted in a severe asthma attack. We planned to perform laryngotracheoesophagoplasty, which provides laryngeal function, but this was precluded by difficulties

controlling her asthma and the need to perform surgery for the DORV as early as possible to ensure long-term survival. After providing sufficient explanation to the parents, a laryngotracheal separation was performed at 1 year 7 months of age to completely prevent aspiration. A vertical incision was made in the cervical region up and down the tracheostomy with the neck in an extended position. An incision was not made into the sternum because we planned to surgically repair the DORV after this procedure. The thyroid isthmus and upper part of the thymus

Table 1
Reported cases of type 4 laryngotracheoesophageal cleft associated with esophageal atresia.

Author	Associated anomalies	Approach for LTEC	Esophageal reconstruction	Airway management	Full oral feeding	Outcome
Ogawa T et al., 1989	Tracheomalacia	Lateral approach	Cervical esophagostomy	Not mentioned	Not achieved	Survive
Ratan SK et al., 2001	None	Not performed	TEF ligation	Intubation	Not achieved	Died a week of age
Mathur NN et al., 2006	Duodenal atresia Hyaline membrane disease, intraventricular hemorrhage	Anterior approach Not performed	Esophagoplasty Not performed	Tracheostomy Intubation	Not achieved Not achieved	Survive Died age 7 d
Our patient	Duodenal atresia, DORV	Separation of the larynx	TEF ligation	End-tracheostomy	Not achieved	Survive

Abbreviations: LTEC, laryngotracheoesophageal cleft; DORV, double outlet right ventricle; TEF, tracheoesophageal fistula.

were dissected in the middle, widely exposing the trachea. The trachea and upper esophageal pouch were taped, and the trachea was then transected at the level of the tracheostomy. The upper esophageal pouch was then separated from the trachea (Fig. 3). The oral side of the esophagus was so short that a cervical esophagostomy could not be established. The larynx was closed in two layers, and an end-tracheotomy was created. To prevent relapse of the fistula, the sternohyoid and sternothyroid muscles were interposed between the trachea and esophagus. No complications developed after surgery, and the severe asthma attack was resolved because of the complete prevention of aspiration. As a radical surgery for her congenital heart disease, intracardiac repair of DORV was performed at the age of 2 years 2 months. Oral cavity secretions are currently expelled from the mouth and nose because the oral side of the esophagus still has a blind end, but she is in excellent general condition and does not require oxygen administration at 2 years 10 months of age. We are considering reconstruction of the esophagus.

2. Discussion

LTEC is an extremely rare congenital malformation. The estimated incidence of LTEC is 1 in 10,000–20,000 births [1]. Various other associated malformations have also been reported, with esophageal atresia occurring in 20–37% of cases and heart malformations in 16–33% of cases [1]. To the best of our knowledge, only four cases of type 4 LTEC with type C esophageal atresia as in our case were reported between 1972 and 2012 (Table 1) [3–5]. Moreover, our report describes the first case of type 4 LTEC associated with type C esophageal atresia, duodenal atresia, and severe congenital heart disease.

To repair the LTEC, various cleft-closing techniques have been described. An endoscopic procedure was recently reported for low-grade LTEC, but high-grade LTEC still requires an open approach. The anterior laryngotracheal approach is most widely accepted [1,6]. The objectives of these procedures are to secure the airway without intubation or tracheostomy and to establish oral feeding without gastrostomy. However, a long time is reportedly required to reach these goals [5–7]. Among the four cases of type 4 LTEC associated with esophageal atresia, one case involved an anterior approach, one involved a lateral approach, and two were inoperable. Only two of these patients survived for more than 1 year, and they were unable to establish oral feeding completely. One of them was dependent on a tracheostomy for more than 10 years. In our case, we performed laryngotracheal separation and established an end-tracheostomy to completely and immediately prevent aspiration. To date, only Hashizume et al. [8] have reported laryngotracheal separation in a case of LTEC. The authors reported that laryngotracheoesophagoplasty was performed with an anterior approach via a full sternotomy for the first time. However, because the fistula reformed and laryngeal function could not be restored following the operation, laryngotracheal separation was performed as a second operation.

Mathur et al. [5] reported that it is possible to access the entire length of the trachea via a cervical “T” incision. In our experience,

the upper esophagus pouch and almost the whole length of the trachea were sufficiently exposed via a cervical approach with the neck in an extended position without a sternotomy. We were also able to easily interpose the sternohyoid and sternothyroid muscles between the separated trachea and esophagus, preventing fistula reformation after laryngotracheal separation as Shino et al. [9] reported. The patient was quite well postoperatively. The asthma attack ceased immediately, and radical surgery for the DORV was performed uneventfully.

3. Conclusion

Ideally, the surgical repair for LTEC should be laryngotracheoesophagoplasty to obtain function of the larynx. However, this procedure might be difficult depending on the severity of the LTEC and the accompanying malformations. We believe that our procedure is a feasible choice for long-term survival in patients with a severe LTEC with congenital heart disease.

Conflicts of interest

There are no relationships, conditions, or circumstances that present a potential conflict of interest.

Sources of funding

There were no sources of funding regarding the present work.

Consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal on request.

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