

Type IV laryngotracheoesophageal cleft repair by a new combination of lateral thoraco-cervical and laryngoscopic approaches

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Abstract Type IV laryngotracheoesophageal cleft (LTEC) is a rare congenital anomaly that is associated with high morbidity and mortality despite various forms of surgical repair. This article presents our strategy for surgical management of type IV LTECs using a combination of lateral thoraco-cervical and laryngoscopic approaches.

Keywords Endoscopic approach · Anterior approach · Children · Laryngotracheoesophageal cleft type IV · Lateral approach

Introduction

Congenital laryngeal anomalies occur in approximately 1 in 2,000 live births, and less than 0.3 % of these are attributable to laryngeal clefts [1]. The severity of the presenting symptoms is dependent on the type of cleft [2]. While a number of classifications of clefts have been based on the distal extent of the cleft [1], the type IV laryngotracheoesophageal cleft (LTEC) in Ryan's classification [3] is the severest form as it extends to one or both of the main-stem bronchi.

Several surgical approaches for treating LTECs have been reported in the literature [1–8], including lateral,

anterior, and endoscopic approaches. One of these approaches is usually selected based on the type of cleft; however, surgical repair of type IV LTECs is extremely demanding and is associated with high morbidity and mortality. A timely diagnosis as well as a more appropriate strategy for repairing type IV LTECs is important for improving the survival and quality of life of patients [2].

Operation

Our method of surgical management of type IV LTECs combines two surgical approaches. The first is a right lateral thoracotomy in combination with a cervical approach [3], and this is used to repair the cleft from the third tracheal ring through to the left main-stem bronchus, while the patient is placed on extracorporeal membrane oxygenation (ECMO). After a tracheostomy the second is a modified endoscopic approach [1], and this is used for repairs from the arytenoids to the second tracheal ring. Anterior approach [4] through the tracheostomy is reserved for any remaining cleft repairs and is used when required.

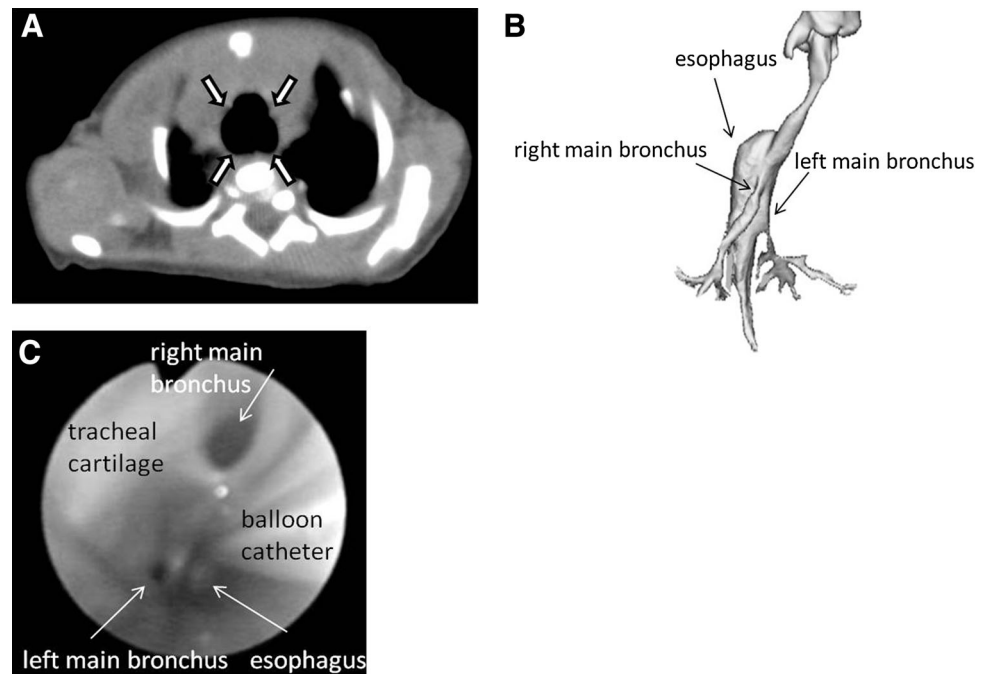
Case report

A male infant was born to a 26-year-old mother at our hospital of 38 weeks gestation weighing 1,948 g. His 1- and 5-min Apgar scores were 4 and 8, respectively. The pregnancy was complicated by polyhydramnios, and a prenatal sonography revealed a fetal esophageal dilatation with an absent fetal stomach. At birth, the infant was noted to have a weak cry, copious secretions, labored breathing, and tachypnea. Emergency computed tomography (CT) confirmed the diagnosis of a type IV LTEC extending to

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Fig. 1 Images of a type IV laryngotracheoesophageal cleft (LTEC). **a** CT image of a type IV LTEC. The arrows indicate the cleft. **b** An image showing the three-dimensional reconstruction of a type IV LTEC. **c** An endoscopic image of the cleft



the left main-stem bronchus (Fig. 1a, b). The infant also had scoliosis and an atrial septal defect.

On day 5, ligation of the abdominal esophagus and gastrostomy were performed to allow enteral feeding while preventing gastroesophageal reflux (GER). The general anesthesia was performed using a cuffed endotracheal tube after clamping esophagogastric junction with a balloon catheter. The endoscopic finding revealed the very wide cleft, and too short and straight tracheal cartilages (Fig. 1c).

On day 54 [bodyweight (BW): 2,900 g], a retropleural approach through the right lateral thorax was used in combination with a right lateral cervical approach to expose the tracheoesophageal groove from the third tracheal ring to the left main-stem bronchus. The more cephalad trachea, pharynx, and right recurrent laryngeal nerve were left intact. With the infant placed on ECMO, the cleft through to the left bronchus was repaired with an esophageal flap created from the left lateral esophageal wall (Fig. 2a). The flap provided a tension-free closure without redundant esophageal tissue and formed a membranous trachea. A custom-designed bifurcated endotracheal tube was used as a tracheal and bronchial stent to secure the airway (Fig. 2b). The remaining esophagus was closed longitudinally with a No. 5 French feeding tube placed inside to act as an esophageal stent and provide drainage. A two-layer closure was performed in both the trachea and esophagus. Because the ECMO cannula was inserted in the right auricle, a long pericardium flap was placed between the tracheal and esophageal closures as a septal patch in the thorax. A portion of the sternothyroid

muscle was placed between the tracheal and esophageal suture lines in the neck. The LTEC above the third tracheal ring was left intact, because an attempt at repair via this lateral approach might have caused a recurrent laryngeal nerve injury and left–right asymmetry of the arytenoids. A tracheostomy was performed on the second tracheal ring while the bifurcated endotracheal tube was removed.

At month 4 (BW: 4,000 g), the remaining cleft across the arytenoids to the second tracheal ring was repaired via a modified endoscopic approach. The laryngoscope introduced into the cleft clearly showed its two lateral borders and provided access to its lower end. Longitudinal dissection of the mucosa (mucosectomy) along both lateral borders of the cleft was performed with forceps from the lower end (above the tracheostomy tube) up to the level of the arytenoid cartilages. The cleft was then closed with a single layer of sutures with juxtaposition of the borders and separated stitches; the knots were tied on the laryngotracheal side (Fig. 2c). Once the infant's respiratory status had improved after the cleft repair, the abdominal esophagus band was untied and fundoplication was performed along with esophageal dilatations for esophageal stricture.

Esophagoscopy and CT revealed a pinhole tracheoesophageal cleft remaining at the level of the tracheostomy (Fig. 3a, b) at month 9 (BW: 5,400 g), and this was closed via a modified anterior approach through the tracheostomy orifice with the infant placed on ECMO. A tracheoesophageal septum was created by two-layer suturing of the esophageal and tracheal mucosal flaps.

At 1 year of age, although the infant is dependent on the tracheostomy due to tracheobronchomalacia, he is being

Fig. 2 a, b
Bronchotracheoplasty with a custom-fitted bifurcated endotracheal tube stent.
c Endoscopic repair of the cleft

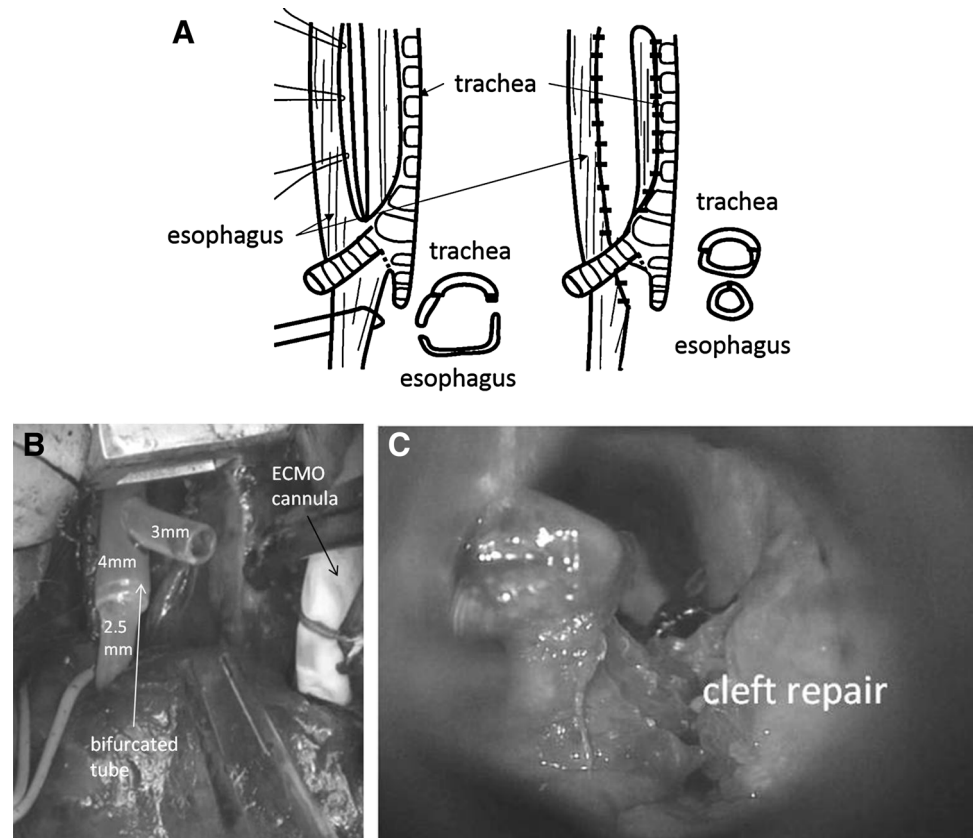
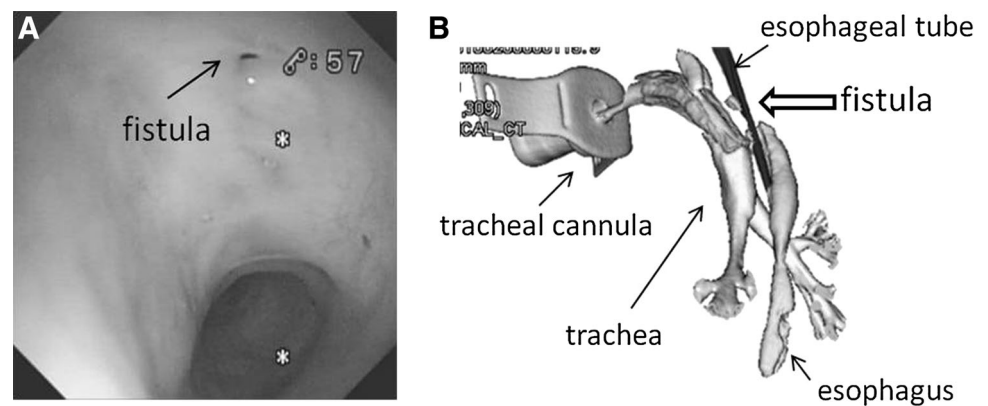


Fig. 3 a Endoscopic image of a residual cleft and fistula.
b Image showing the three-dimensional reconstruction of the residual cleft and fistula



weaned from ventilator support, and appears to be cognitively normal. In addition, his laryngeal function may be preserved because he had no history of dysphagia and tolerates oral feeding.

Discussion

Since LTECs are very rare congenital anomalies, no single institution has sufficient experience to propose a convincing standard algorithm for surgical treatment. The prognosis for type IV LTECs in particular remains poor despite

recent advances in surgical techniques and postoperative care [2]. The indication for surgery depends on the overall status of the patient [4].

Three major surgical approaches for LTEC repair have been reported as follows: lateral [2, 3, 5, 6], anterior [4, 7], and endoscopic [1, 8]. The advantages and disadvantages of each approach have also been discussed [1–8].

The lateral thoracic approach provides excellent access to the lower portion of the LTEC when there is sufficient esophageal wall available for donating tissue to close the trachea, while retaining enough tissue to close the esophagus without stricture [3, 6]. However, the lateral approach

cannot be used for precise laryngeal repair. It also poses a risk of recurrent laryngeal nerve injury and fistula formation [7], although the interposition of bulky vascularized tissue has helped to decrease the incidence of fistula formation [3, 5].

The main advantage of the anterior approach is that it provides excellent direct operative access with minimal dissection at the cervical level and there is no risk of recurrent nerve damage [4, 7]. However, anterior midline incision of the larynx and trachea can have a negative impact on the stability of the airway [1]. Moreover, when viewed through the anterior approach, it is difficult to judge how much of the pharyngeal or esophageal mucosa should be used for a reconstruction without binding [1]. Placement of a posterior cartilage graft between the arytenoids may be necessary [7].

The endoscopic approach provides the surgeon with a permanent axial view of the airway and is useful for avoiding the inclusion of excess mucosa in the reconstructed airway, which can potentially lead to laryngotracheal stenosis. Additionally, this approach preserves the best possible vascular supply to the mucosa used for the repair [1, 8]. However, the surgical indication for an endoscopic approach for LTEC is limited to patients with no involvement of the tracheal rings [8].

Considering the advantages and disadvantages of these three approaches, we propose that a combination of lateral thoraco-cervical approach and endoscopic approach should be used for clefts extending to the carina and beyond. Especially, in our case whose cleft was very wide and tracheal cartilages were too short, it would be really difficult to repair the cleft and at a high risk of fistula formation by an anterior approach. In our case, the first approach (a right lateral thoracotomy in combination with a cervical approach) provided far superior access for surgical repair of the type IV cleft extending from the third tracheal ring through to the left main-stem bronchus. Considering the risk of recurrent nerve injury, the more cephalad trachea and pharynx were left intact. The subsequent modified endoscopic approach enabled the repair of the remaining cleft from the arytenoids to the second tracheal ring as well as reconstruction of the larynx with minimal invasion. In addition, one staged repair by lateral and endoscopic approaches would be thought too invasive for this low-birth weight infant with type IV LTEC. Although a small cleft remained at the level of the tracheostomy in our case, we suggest that the combination of these two approaches can enable complete repair of type IV LTECs; an anterior approach via a tracheostomy orifice is suitable for closing any remaining small cleft.

Patients who survive the initial repair of a type IV LTEC face a difficult postoperative course. Tracheomalacia is a

major postoperative challenge [2]. After repair of a cleft extending to the carina or beyond, the “esophageal portion” of the airway offers less stability than a normal airway. There may also be weakness or deformity of the tracheal rings [5]. Another common problem is GER. Controlling GER is necessary to decrease the risk of repair breakdown and fistula formation. Furthermore, patients typically continue to suffer from chronic lung disease secondary to recurrent aspiration [2].

In our case, although follow-up period is short, tracheobronchomalacia remains a problem and a tracheostomy is still required; however, tracheomalacia has been reported to improve over time because both the rigidity of the tracheal rings and the size of the tracheal lumen increase as the patient grows, decreasing the propensity for tracheal collapse [2]. To prevent GER, ligation of the abdominal esophagus and subsequently fundoplication to remove the band of the esophagus are useful.

In conclusion, our operative strategy for type IV LTECs involves a combination of lateral thoraco-cervical and laryngoscopic approaches and is effective for cleft repair. The strategy facilitates the preservation of laryngeal function while preventing fistula formation.

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References

1. Sandu K, Monnier P (2006) Endoscopic laryngotracheal cleft repair without tracheostomy or intubation. *Laryngoscope* 116:630–634
2. Owusu JA, Sidman JD, Anderson GF et al (2011) Type IV laryngotracheoesophageal cleft: report of long-term survivor successfully decannulated. *Int J Pediatr Otorhinolaryngol* 75:1207–1209
3. Ryan DP, Muehrcke DD, Doody DP et al (1991) Laryngotracheoesophageal cleft (type IV): management and repair of lesions beyond the carina. *J Pediatr Surg* 26:962–970
4. Moukheiber AK, Camboulives J, Guys JM et al (2002) Repair of a type IV laryngotracheoesophageal cleft with cardiopulmonary bypass. *Ann Otol Rhinol Laryngol* 111:1076–1080
5. Kawaguchi AL, Donahoe PK, Ryan DP (2005) Management and long-term follow-up of patients with types III and IV laryngotracheoesophageal clefts. *J Pediatr Surg* 40:158–165
6. Donahoe PK, Gee PE (1984) Complete laryngotracheoesophageal cleft: management and repair. *J Pediatr Surg* 19:143–148
7. Geller K, Kim Y, Koemple J et al (2010) Surgical management of type III and IV laryngotracheoesophageal clefts: the three-layered approach. *Int J Pediatr Otorhinolaryngol* 74:652–657
8. Rahbar R, Rouillon I, Roger G et al (2006) The presentation and management of laryngeal cleft: a 10-year experience. *Arch Otolaryngol Head Neck Surg* 132:1335–1341