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Case report

# Laryngotracheoesophageal Cleft Type 3 and Severe Laryngotracheomalacia; Delayed Surgical Repair, a Treatment Challenge with an Excellent Outcome

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ABSTRACT

Laryngotracheoesophageal clefts (LTEC) are rare malformations which involve the upper respiratory and digestive tract. Surgical repair should be undertaken promptly to maintain a secure airway and prevent serious pulmonary aspiration. This paper reports the first case of LTEC type 3 with severe laryngotracheomalacia that was brought to Mofid children's hospital in late infancy with a poor health status. Delayed defect correction was our team strategy for the patient when she had achieved good weight gain. At the age of 22 months in collaboration with the pediatric surgical and otolaryngologist team, the repair of the laryngeal cleft was done with lateral open approach method. She was discharged with tracheostomy and gastrostomy. In the next six months follow up after the surgery tracheostomy decannulation and gastrostomy tube removal were done and the infant is now in regular follow-up.

## 1. Introduction

Laryngotracheoesophageal clefts (LTEC) are uncommon inherent abnormalities that result secondary to defect in fusion of the tracheoesophageal wall during embryological development [1]. The reported incidence is variable from 1 in 10,000 to 1 in 20,000 live births, with more occurrence in males. Most cases are sporadic while the few others are associated with syndromes such as Opitz-Frias or Pallister-Hall, or congenital anomalies like tracheoesophageal fistulas [2]. Patients who are diagnosed as a case of laryngeal clefts develop disorders related to swallowing and airway, leading to recurrent aspiration pneumonia, labored breathing, and failure to thrive [1]. Therefore, a proper and well-timed diagnosis has a great importance. As there are no pathognomonic symptoms and signs for this rare congenital anomaly and the presenting features can mimic a variety of other clinical conditions, such as hyperreactive airways, laryngomalacia, gastroesophageal reflux, and neuromuscular swallowing disorders, therefore diagnosis is hardly possible without a high index of suspicion besides utilizing bronchoscopic evaluation [3]. We report the first case of LTEC managed by pediatric surgery, otolaryngology and pulmonology team of MOFID children's hospital.

## 2. Case presentation

A 45-day old female infant presented with recurrent episodes of nasal regurgitation, severe choking and respiratory distress with a history of several times admissions in different hospitals. The child was full term, first in birth order and delivered by normal delivery. The infant was the outcome of consanguineous marriage. The prenatal or natal history was insignificant. After birth, the child developed noisy breathing, difficulty and choking episodes while being fed. The child was investigated with necessary investigations and keeping suspicion of tracheoesophageal fistula (TEF), barium swallow study was done. The study showed leakage of the contrast dye into the trachea. It was followed by rigid bronchoscopy and diagnosis of laryngo-tracheoesophagial cleft type 1 with significant laryngomalacia was made. The patient was discharged with anti-reflux treatment. As the infant developed poor weight gain, therefore gastrostomy tube was inserted at the age of 8 months. She was first time brought to our center at the age of 13 months when she was admitted to pediatric surgery ward and opinion was taken from pediatric pulmonology department and fiberoptic bronchoscopy was planned. A posterior LTEC was seen during fiberoptic bronchoscopy that extended down distally to the 3rd tracheal cartilage (Figs. 1 and 2). There was associated severe

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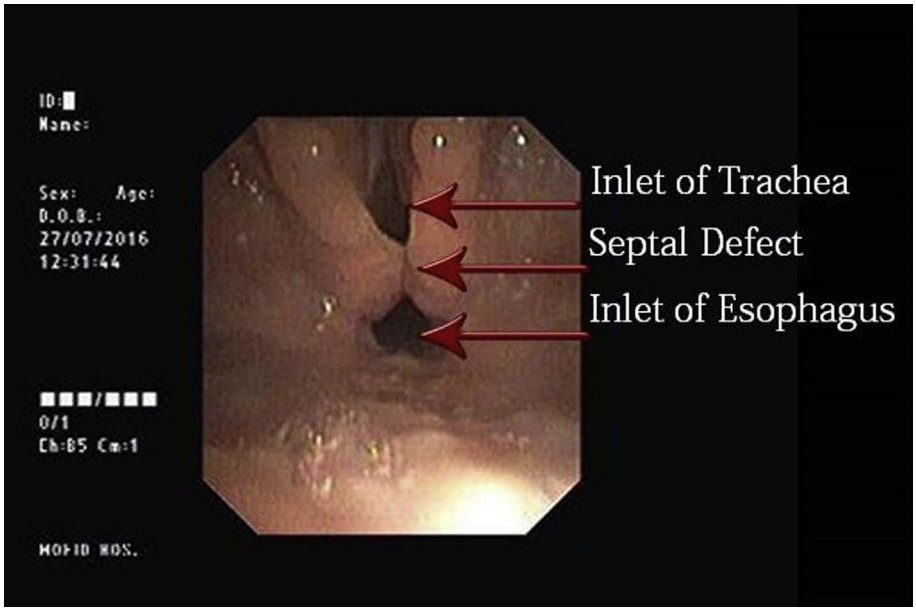


**Fig. 1.** LTEC (proximal view).

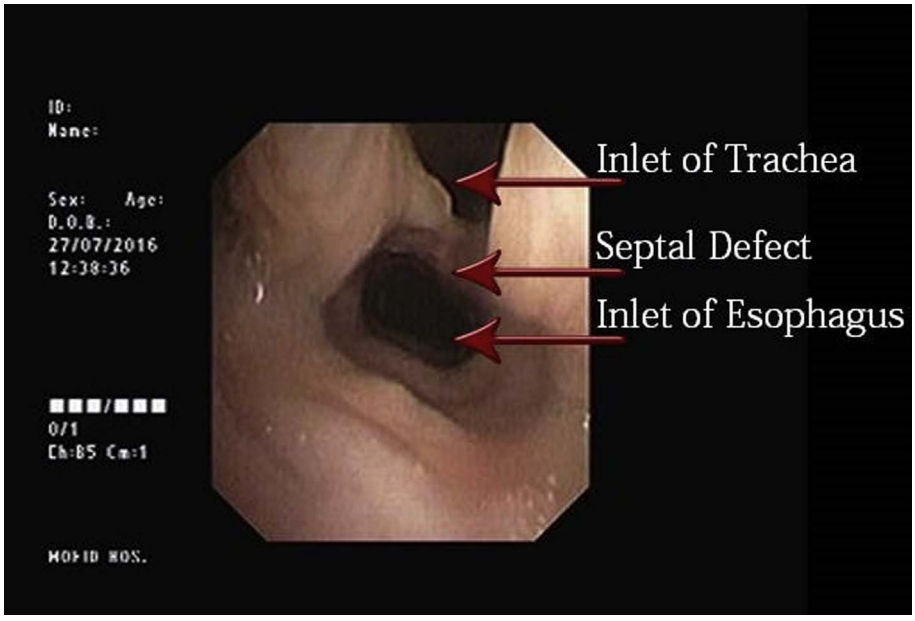


**Fig. 2.** LTEC (distal view).

laryngotracheomalacia (Fig. 3) with no other airway anomaly. As the patient was emaciated with poor weight gain (BMI = 11), so the total correction was postponed for the next few months so that she can achieve good weight gain.

At the age of 22 months in collaboration with the pediatric surgical and otolaryngologist team, the repair of the laryngeal cleft was done with lateral open approach method. Under general anesthesia and in a sterile setting, the operation started with a transverse incision in the right lateral side of the neck. Then sternocleidomastoid muscle was pushed aside, so the thyroid, arytenoid and cricoid cartilages in the right side became visible. Pyriform sinus was dissected from the lower end of the thyroid cartilage. True vocal cords, trachea, and esophagus were defined. A nasogastric tube was passed through the esophagus. Both trachea and esophagus were repaired with separate sutures.

Finally, from anterior neck approach,2 cm upper to sternal notch, tracheostomy tube was placed for the patient. The postoperative course was insignificant and she was discharged to home after 2 weeks (Fig. 4). Tracheostomy decannulation was done after 2 months of operation. During the next 6 months, follow-ups after surgery the patient had good weight gain and gastrostomy tube was also removed and the site was closed.

## 3. Discussion

Laryngotracheoesophageal cleft (LTEC) is a congenital midline anomaly, which defines when there is a defect of the posterior part of larynx and trachea and the anterior wall of the esophagus. It is rare but attributed with considerable mortalities and morbidities [1,6]. There is no specific pattern of inheritance and most reported cases had a sporadic pattern, however, some reports from familial occurrence with autosomal dominant pattern also have been reported in the literature [1].

Respiratory bud develops from diverticulum on foregut, at the 4th week of gestation. Tracheobronchial groove starts to exist on both sides and reach together in the midline and form the tracheoesophageal septum by the 5th week of gestation. Failure in development of tracheoesophageal septum results in the occurrence of LTEC. In which phase the fusion

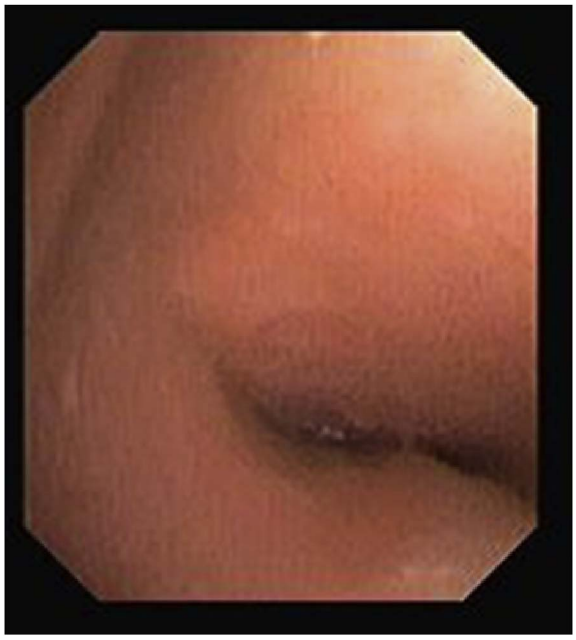

**Fig. 3.** Severe tracheomalacia.

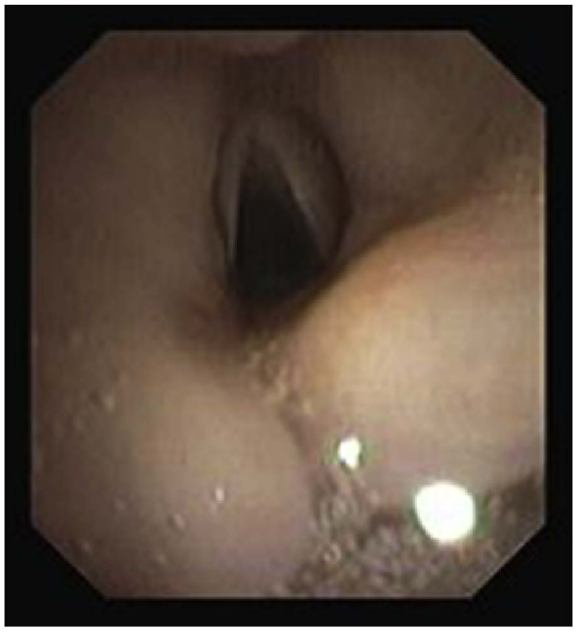

**Fig. 4.** Post surgical repair.

process interruption occurs, determine the length of cleft [3,7]. Males are affected more than females [1,8], but in the index case, we report a female in contrast to most of the earlier reported cases. Contributing conditions such as prematurity and polyhydramnios have been reported in the majority of previous cases [8], but surprisingly our case was born at term without a prenatal history of polyhydramnios.

The Benjamin-Inglis classification system is the most clinically used classification for laryngeal clefts. It was published in 1989 and according to it, LTECs are categorized into four types [4]. Type I defined as a defect of inter-arytenoid to the level of the true vocal folds, type II is when the defect affects the posterior cricoid cartilage, type III is the extension of the defect entirely through the posterior cricoid cartilage and possible involvement of the cervical trachea, and type IV defined as an extension into the intrathoracic trachea (4). Among these types, particularly types 3 and 4 are a real challenge in diagnosis and clinical management [5] and these patients need multidisciplinary approach from pediatric surgeons, otolaryngologists, pediatric pulmonologists, and nutritionists.

Tracheoesophageal fistula, anal atresia, cleft lip, Meckel's diverticulum, bronchial and tracheal stenosis and cardiovascular defects, may occur with LTEC. These anomalies have an influence on the clinical course and prognosis of the affected patient [1,6]. The index case did not fit any specific syndrome, except severe laryngotracheomalacia, there were no other congenital anomalies. Association of LTEC type 3 with severe tracheobronchomalacia has been reported by Mitchell DB et al. [9] but till date, no case of LTEC type 3 and severe laryngotracheomalacia with no other congenital anomaly has been reported other than the index case. One of the challenges faced by our team in the management of the case was to decide whether to operate her immediately after the proper diagnosis was made or not. Despite knowing that the timing and approach for surgical repair depends on the severity of symptoms, associated abnormalities, and the type of cleft and in most of the cases early diagnosis and surgical repair of laryngeal cleft is practiced to reduce the irreversible pulmonary damage and other associated morbidities that may occur as a result of recurrent aspiration [3], but in the index case the exact diagnosis was made at the age of 13 months, and according to pediatric surgeon team, good calorie intake and good health status were the priority over the surgical correction of the cleft and with close follow-up of the patient, therefore the surgery was postponed till health status of the patient became more acceptable. This was the first experience of our center for such type of cases with an excellent outcome.

## 4. Conclusion

Considering this point that LTEC and laryngeal clefts are rare anomalies lack of practical experience in some centers may cause a delay in appropriate diagnosis. We report the first case of LTEC type 3 from our country which was diagnosed after infancy and despite the delayed correction strategy, she has survived well.

## Conflicts of interest

The authors report no conflicts of interest for this article.

## Appendix A. Supplementary data

Supplementary data related to this article can be found at http://dx.doi.org/10.1016/j.rmcr.2017.10.007.

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