Case report
Anesthesia management of a patient with a laryngotracheo-esophageal cleft

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Summary
Laryngotracheo-esophageal cleft is a rare congenital anomaly that results from complete or partial failure of the development of the tracheoesophageal septum. The presenting symptoms include stridor, respiratory distress, and coughing or cyanotic episodes with feeding. There are four classifications for laryngeal clefts; the severity depends on the type present. We discuss the anesthesia management of a neonate with a Type IV cleft who presented for an emergency gastric division to prevent pulmonary aspiration and later returned for final repair of the defect.

Keywords: congenital airway anomaly; laryngeal cleft defect; laryngotracheo-esophageal cleft

Introduction
The trachea and esophagus originate from one common tube and are separated by the rostral development of the tracheoesophageal septum, which is complete at approximately 35 days of gestation (1). Complete or partial failure of the septum to develop results in a laryngotracheoesophageal (LTE) cleft. Laryngeal clefts occur with varying degrees of severity: the mildest form (Type I) is almost asymptomatic, and the most severe forms are life threatening. These anomalies are rare and may be difficult to diagnose because of their relative obscurity.

There have been a few classifications for laryngeal cleft defects proposed. The first was described by Petterson in 1955 (2,3) and was later modified by Evans (4). In this modification, Type I is a supraglottic interarytenoid cleft located above the vocal cords and not involving the cricoid lamina, Type II includes a posterior defect of the cricoid cartilage and extends to the cervical trachea, and Type III is a defect that extends to the thoracic trachea. In 1991, Ryan et al. (5) described three children with a more extensive defect extending beyond the carina into either one or both main stem bronchi. This more extensive defect is classified as Type IV (5) (Figure 1).

Presenting symptoms include stridor, excessive secretions, coughing and/or cyanosis with feeding, recurrent pneumonia, and respiratory distress. Type I defects may remain undetected, and the patient may present as an adult with minor symptoms such as hoarseness and recurrent infections (6). More severe defects present with symptoms during the early neonatal period, and the diagnosis is often...
presumed to be tracheoesophageal fistula. We present a neonate diagnosed with Type IV LTE cleft and discuss the anesthesia management as well as the available literature.

Case report

A 1-day-old Caucasian female infant was transferred to our institution from a community hospital. The baby was born at 36-week gestation to a 25-year-old gravida 3 para 1 female. A fetal ultrasound at 20-week gestation was unable to visualize the fetal stomach and esophageal atresia without fistula was suspected. The pregnancy was complicated by worsening polyhydramnios. Labor commenced after spontaneous rupture of membranes, and the infant was delivered vaginally. Birth weight was 2.55 kg with Apgar scores of 7 at 1 min and 9 at 5 min. The infant was bag-mask ventilated briefly at birth because of respiratory depression and then was supported with nasal continuous positive airway pressure (CPAP) ventilation.

On admission to our neonatal intensive care unit, the infant showed signs of respiratory distress with mild retractions and marked oropharyngeal secretions. Several attempts were made to intubate the trachea. On each attempt, the tracheal tube seemed to be placed into the trachea with audible bilateral breath sounds. However, the baby continued to make crying noises after tracheal tube placement and air entry could be heard over the stomach with positive pressure ventilation. The baby was subsequently extubated and laryngoscopy and bronchoscopy were scheduled to establish a diagnosis.

The infant underwent laryngoscopy and bronchoscopy in the operating room while awake and breathing spontaneously. Laryngoscopy demonstrated a complete absence of the posterior tracheal wall to a level immediately above the carina. The left main bronchus was abnormal with a small posterior opening and a larger anterior opening. Vocal cord motion was not assessed. The diagnosis of LTE cleft Type IV was established, and the baby was returned to the neonatal intensive care unit in stable condition, breathing spontaneously through her natural airway.

Later the same day, the infant was returned to the operating room for gastric division and gastrostomy tube placement. Physical exam revealed a neonate

![Figure 1](https://example.com/image.png)

Types of laryngeal cleft defects. (Used with permission from Ref. (5).)
with coarse bilateral breath sounds and stridor with crying. Oxygen saturation was 100% on room air. The rest of the examination was unremarkable and showed no other obvious malformations. In the operating room, standard monitors were applied including the ECG, pulse oximetry, and NIBP and an existing umbilical artery catheter was transduced. After discussion with the surgeon and otorhynologist, the tracheal tube was placed through the esophageal orifice. This placement allowed for the insertion of a larger cuffed tube that would permit suctioning and better control of secretions. Laryngoscopy was performed with the infant awake, and a 4.0-cuffed (TT) was inserted into the esophageal orifice. Bilateral breath sounds were heard and endtidal CO\textsubscript{2} was present. General anesthesia was induced with incremental increases of sevoflurane to 2% with the patient breathing spontaneously. Ketamine hydrochloride was also administered in 1-mg increments as surgery commenced, for a total of 3 mg. An upper abdominal incision was made and proximal gastric division was performed, with placement of a gastrostomy tube in the distal portion of the stomach. The stomach was extremely small, measuring approximately 5-cm length and 3 cm at its greatest width.

During the procedure, the infant required assisted ventilation for periods of hypoventilation, which was performed with gentle hand ventilation. The endtidal CO\textsubscript{2} waveform was lost intermittently throughout the procedure, although bilateral breath sounds were audible. The absence of endtidal CO\textsubscript{2} was attributed to a large leak through the glottic opening. Blood gas analysis showed pH 7.36, PaO\textsubscript{2} 19 kPa (147 mm Hg), pCO\textsubscript{2} 3.9 kPa (30 mm Hg), HCO\textsubscript{3} 17 mmol\textpermm\textsuperscript{-1}, and oxygen saturation 100%. SpO\textsubscript{2} was 97–100% throughout the procedure. At the conclusion of the procedure, the baby was awakened and extubated. After a few minutes of unassisted respirations, the baby showed signs of respiratory distress with suprasternal and abdominal retractions present. This distress was alleviated with 5–10 cm CPAP applied through a facemask. She was reintubated with a 4.0-cuffed tracheal tube inserted through the esophageal orifice. The baby was allowed to breathe spontaneously with 5–10 cm CPAP applied via the Mapleson circuit during the transfer to the intensive care unit. The infant was transferred to the unit in stable condition.

She was extubated on postoperative day 4 and was fed through the gastrostomy tube; the plan was to allow her to grow before proceeding with the definitive repair. The ultimate goal was for the baby to reach a weight of 5 kg. The infant was reintubated on three occasions following the gastric procedure over a period of 5 weeks for recurrent aspiration pneumonia. As a result of the multiple episodes of aspiration pneumonia, the decision was made to proceed with the definitive repair of the LTE cleft.

The child was taken to the operating room for this procedure at age 7 weeks with a weight of 3.7 kg. Induction was with isoflurane through an already present tracheal tube. She was reintubated with a 5.0-cuffed endotracheal tube (ETT) placed through the esophageal orifice, and the isoflurane was discontinued. We gave a remifentanil infusion throughout the procedure, and neuromuscular blockade with pancuronium and vecuronium. A sternotomy was performed, and exposure of the larynx and tracheoesophagus obtained. Cardiopulmonary bypass (CPB) was then initiated following the placement of appropriate arterial and venous cannulae and administration of heparin. The trachea and esophagus were divided, and the strap muscle interposed between the structures. At the completion of the airway and tracheal reconstruction, she was separated from CPB.

Prior to initiation of bypass, a bronchoscopy was performed that showed the left main bronchus entering the trachea as two smaller diameter orifices rather than a single lumen. At the conclusion of the reconstruction procedure, the left lung did not inflate sufficiently, and the left bronchus was found to be edematous but patent. She remained intubated and was taken to the Pediatric Intensive Care Unit (PICU), where mechanical ventilation was instituted. Following the repair, the infant had problems with continued aspiration of pooled secretions as well as severe tracheomalacia. She had several bronchoscopies as well as two failed attempts at tracheostomy because of difficulty ventilating during the attempts and difficulty placing a tracheostomy tube. She had a tracheostomy at the third attempt. During her hospital stay, she also had a Ladd’s procedure, Roux-en-Y, and re-anastomosis of the distal esophagus and stomach at 5 months of age. She tolerated these procedures well.
She had been discharged home after spending a total of 101 consecutive days in hospital since birth. At the time of discharge, she had a 4.0-cuffless tracheostomy tube and was on mechanical ventilation. Her weight at discharge was 6.8 kg, which was in the 10th to 21st percentile range for her age.

Discussion

The lower respiratory tract begins to develop at approximately 4-week gestation from a midline laryngotracheal groove that forms in the ventral portion of the foregut (7). By the 5th week, the laryngotracheal groove is transformed into a laryngotracheal diverticulum (8) that grows parallel to the foregut. Tracheoesophageal folds then form to create the laryngotracheal septum, which separates the laryngotracheal diverticulum from the foregut. This results in the creation of the primordial esophagus and the laryngotracheal tube, which develops caudally into the trachea, bronchi, and distal airways. The laryngotracheal septum forms in a caudal-to-rostral direction and is usually complete by 35-day gestation (7,8).

Presumably, disruption in the cephalad development of the laryngotracheal septum results in a laryngeal cleft defect. It has been proposed also by Merei and Hutson (8) that an LTE cleft may occur as a result of failure of the laryngotracheal diverticulum to form from the foregut. There are no definitive contributing factors associated with these defects, although adriamycin has been reported to induce esophageal atresia and tracheoesophageal fistula in fetal rats (9). LTE defects may be associated with other congenital anomalies and have been described in two syndromes: Opitz–Frias (or G syndrome) and Pallister–Hall syndrome (2,10). Opitz–Frias syndrome includes hypertelorism, cleft lip/palate, cleft larynx, and hypospadias. Pallister–Hall syndrome is associated with hypothalamic hamartoblastoma, hypopituitarism, imperforate anus, postaxial polydactyly, and cleft larynx. Laryngeal clefts have also been found in patients with VATER association, and patients with laryngeal cleft defects may have a coexisting tracheoesophageal fistula as well.

In review of the anesthesia literature, the most recent case report of a patient with laryngeal cleft defect was in a Japanese anesthesiology journal (11). This report describes the incidental finding of LTE cleft in an infant brought to the operating room for repair of presumed tracheoesophageal fistula. Prior to that report, Armitage described three cases of the defect and anesthesia management in 1984 (6). Two of the cases were a mother and son; the son had a Type III defect and died 2 weeks after birth. The mother had a history of persistent hoarseness and recurrent respiratory infections and was found to have a Type I defect that was not diagnosed until after her son had died and her symptoms were further investigated. The third patient in the report was taken to the operating room for ligation of a tracheoesophageal fistula and esophageal atresia repair and then developed a leak around the tracheal tube with intermittent-positive pressure ventilation. Subsequent laryngoscopy revealed a cleft larynx, and the infant died 11 days after birth. The specific anesthesia management for these three cases was not discussed in the report. There are also several case reports in the otolaryngology literature, but these do not specifically address the anesthesia management for this type of defect.

In the current case, we chose to insert the tracheal tube through the esophageal orifice as there was an open communication between the trachea and esophagus. This allowed us to use a larger tube, so we would have better ability to suction secretions. During the gastric division procedure, the baby was allowed to breathe spontaneously to avoid both the distension of the stomach before gastric division and excessive pressure on the proximal gastric suture line after division. Ketamine was administered for analgesia and narcotics were avoided to maintain the respiratory drive. Detection of endtidal CO$_2$ was lost intermittently during the procedure, which corresponded with assisted respirations and was attributed to a significant air leak through the glottic opening and around the tracheal tube.

A crucial step in management of laryngeal cleft defects is establishing a diagnosis. Fortunately, for this infant, her diagnosis was established soon after birth and a gastric division was promptly performed to prevent recurrent aspiration and to allow distal gastrostomy tube feeding. Definitive repair of the defect involves dividing the trachea and esophagus through an anterior sternotomy approach or a transthoracic approach through a thoracotomy incision (5). Maintaining adequate ventilatory support for the repair is challenging, and CPB has been used.
in the past, as well as in this case. Ryan et al. (5) described three patients with a Type IV defect in whom they used a custom-designed bifurcated tracheal tube that allowed for optimal exposure without necessitating CPB. Postoperatively, the patients required prolonged ventilatory support because of the posterior tracheal wall defect and tracheobronchomalacia. Microgastria was present in all three patients with Type IV cleft, as well as in the patient we describe here. Other problems that may occur postoperatively include the occurrence of fistulas and recurrent aspiration once the infant resumes feeding. Only one of three patients described in Ryan et al.’s report survived after cleft repair. The other two died at 8 and 16 months following the repair from unexpected massive aspiration and septic shock, respectively. Horimoto and Yoshizawa described the use of one-lung ventilation with left endobronchial intubation for surgical repair of LTE cleft via a right thoracotomy (12). This was performed successfully, although the authors conceded to problems encountered intraoperatively when the tube was dislodged. The disadvantages in this approach include difficulty placing the tracheal tube into the bronchus, use of a tube that may dislodge more easily, and reliance on one-lung ventilation for a prolonged procedure.

The mortality rate for an LTE cleft increases with severity, ranging from approximately 43% in Types II and III to 90% for Type IV (13). Patients with these defects require early diagnosis and intervention to prevent significant pulmonary damage from aspiration. A multidisciplinary team approach and effective communication between team members are essential in the management of these patients.

In summary, we describe the successful anesthesia management of an infant with Type IV LTE cleft and discuss the pertinent issues for management of a patient with this type of defect.

References

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