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Endoscopic repair of laryngeal cleft

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Laryngeal cleft (LC) is a rare congenital malformation of the posterior larynx. Diagnosis requires a high index of suspicion because type 1 clefts, which are the most common, can be easily overlooked by the endoscopist. Timely diagnosis and appropriate treatment of LC are essential to prevent permanent pulmonary injury. Presenting symptoms include aspiration, cough, recurrent pneumonia, and dysphagia, with a wide range of severity. A trial of feeding therapy remains the management of choice for type 1 LC. When conservative measures fail, the next step in management is surgical repair. Endoscopic repair of LC is a well-tolerated and a reliable approach that considerably reduces perioperative and postoperative morbidity in type 1, type 2, and select type 3 LCs. In this article, we describe the endoscopic management of LCs, highlighting surgical pearls necessary for success.
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Introduction

Laryngeal clefts (LCs) are congenital midline defects, which are characterized by failure of the posterior aspect of the larynx to fuse. The incidence of LC is reported to be 1 in 10,000-20,000 live births,[1] and it is slightly more common in boys than in girls with a ratio of 5:3.[2] The diagnosis of LCs, especially type 1 and type 2 clefts has increased recently because of the increased awareness of small LCs.[3]

The Benjamin and Inglis classification is the most commonly used system for describing LCs.[4] According to this classification system, LC type 1 includes supraglottic interarytenoid defects that extend no further caudally than the true vocal folds. Type 2 LCs extend below the level of the true vocal folds to partially involve the cricoid lamina. Type 3 LCs extend completely through the cricoid cartilage with or without extension into the cervical trachea. Type 4 LCs denote the involvement of the posterior wall of the thoracic trachea with extension as far as the carina.

Presenting signs and symptoms of LC are most often respiratory in nature. Chronic cough is common, especially with feeding. Other frequent signs and symptoms include aspiration, congestion, recurrent pneumonia, and difficulty in feeding. The most imperative factor in the diagnosis is considering this uncommon anomaly in the differential diagnosis. The cornerstone of diagnosis is direct laryngoscopy with palpation of the interarytenoid area (Figure 1). Although type 3 and type 4 LCs are more likely to present within the first days of life, a diagnosis of type 1 and type 2 LCs requires a high level of suspicion because of the nonspecific symptoms.

Conservative treatment is attempted first for patients with type I LCs. The treatment includes feeding therapy, gastroesophageal reflux management, and optimization of respiratory status. In patients with continued aspiration or recurrent pneumonia despite conservative management, endoscopic repair can be considered. In this article, we describe the endoscopic management of LCs and emphasize the surgical pearls necessary for success.

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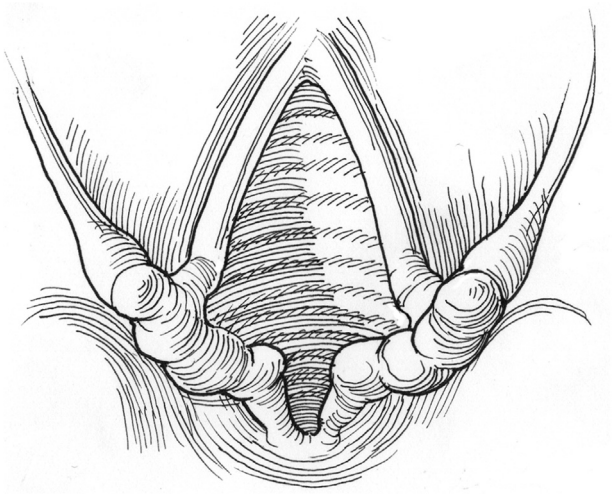

Figure 1 Sketch depicting a type 2 laryngeal cleft extending below the level of the vocal folds to the level of the superior cricoid cartilage.

Indications

Reasons for endoscopic repair of LCs include (1) an insufficient response to medical management and feeding therapy, (2) persistence of aspiration during modified barium swallow study or fiberoptic endoscopic evaluation of swallowing, (3) findings indicative of chronic aspiration on chest imagination with X-ray or computed tomography, and (4) presence of reduced pulmonary status with repeated admissions due to aspiration-related pneumonias.[5-7] Relative contraindications for a successful endoscopic repair include (1) presence of supraglottic airway obstruction that may worsen following endoscopic LC repair and (2) limited access or visualization of the posterior glottis during laryngoscopy.

Surgical technique

Anesthesia

During endoscopic LC surgery, spontaneous ventilation without an endotracheal tube enables clear visualization of the larynx and LC, enables access to the airway for instrumentation, and eliminates the risk of possible damage to the suture line by the endotracheal tube. Working with a skilled anesthetist who has experience in pediatric airway surgeries might help to increase the success of the LC surgery by managing the appropriate anesthetic level for spontaneous ventilation throughout the surgery. Oxygen can be delivered using a side port connected to the laryngoscope, through a Venturi needle placed within the lumen of the laryngoscope or a modified cutoff endotracheal tube at the side of the laryngoscope.

The induction of anesthesia is best achieved by inhalation of sevoflurane or nitrous oxide. The level of anesthesia is maintained with intravenous agents, such as propofol or remifentanil. For analgesia, we prefer to apply topical 4% (1% and 2% are insufficient) lidocaine to the surgical area.

Endoscopic technique

Once an adequate level of anesthesia is achieved, the following steps are necessary for the endoscopic repair of LC:

(1) *Patient position:* A shoulder roll will allow for mild neck extension in the supine position.
(2) *Examination of the entire upper airway:* To confirm the diagnosis of LC and to identify any secondary airway

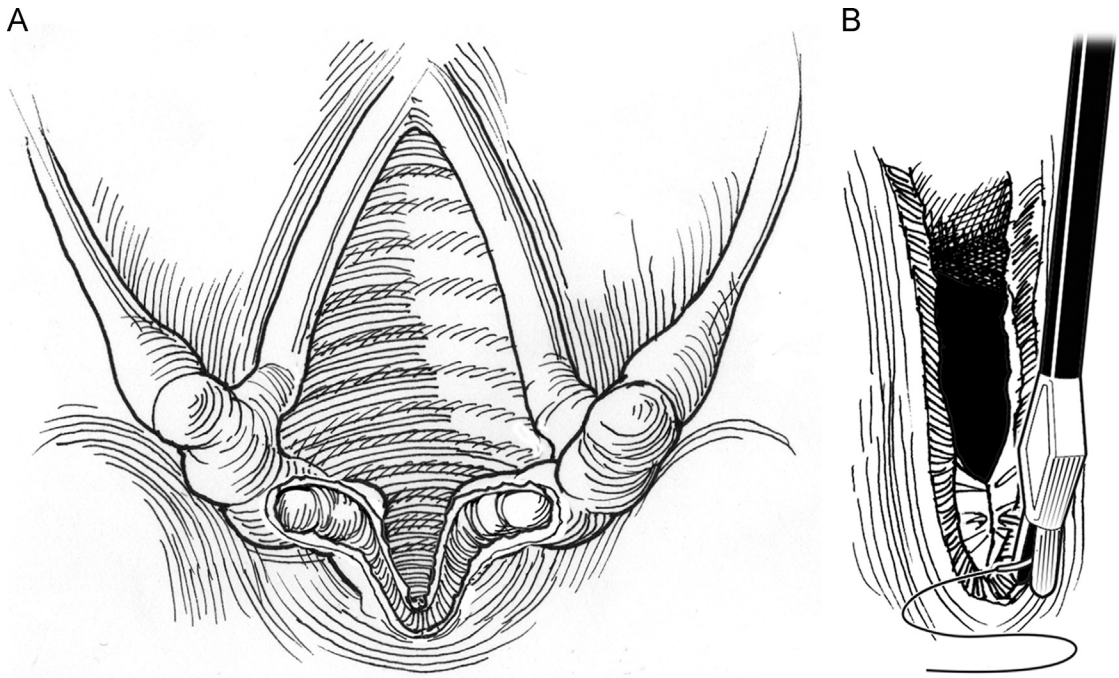

Figure 2 (A) Laryngeal cleft following CO_2 laser ablation of the cleft mucosa. Note that the mucosa at the apex of the cleft is completely ablated to avoid fistula formation. (B) Interrupted suture placement beginning at the apex of the cleft.

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