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Laryngotracheoesophageal clefts

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ABSTRACT

Laryngotracheoesophageal clefts are rare congenital anomalies of the aerodigestive tract. Patients may present with airway and/or swallowing impairments. An approach to evaluation and management is presented. Important pearls for conservative and surgical management are discussed. Open versus endoscopic surgical techniques are reviewed.

Introduction

Laryngotracheoesophageal clefts are rare congenital anomalies that occur during embryological development and result from failure of fusion of the tracheoesophageal septum or lateral growth centers of the posterior-cricoid cartilage.[1–3] There is a male predominance and the annual incidence ranges from 1 in 10,000 to 1 in 20,000 live births.[4] Most cases are sporadic while others are associated with congenital anomalies such as tracheoesophageal fistulas or syndromes such as Opitz-Frias or Pallister-Hall[5]. Patients with laryngotracheoesophageal clefts may present with airway and/or swallowing impairments. These impairments can lead to respiratory distress, recurrent aspiration pneumonia, or failure to thrive.[1–3] Appropriate diagnosis is paramount given the possibility of these concerning sequelae. A comprehensive differential diagnosis should also be considered; the most common diagnoses include laryngomalacia, gastroesophageal reflux, reactive airway disease, and neuromuscular swallowing disorders.

The Benjamin–Inglis classification system published in 1989 describes 4 types of laryngeal clefts.[6] Type I involves an interarytenoid defect to the level of the true vocal folds, type II is partial extension through the posterior cricoid cartilage, type III is an extension completely through the posterior cricoid cartilage and possible extension into the cervical trachea (Figure 1A), and type IV involves extension into the intrathoracic trachea (Figure 1B).

Diagnosis

Evaluation for the diagnosis of laryngeal clefts is variable.[1–3,7–10] Most diagnostic practices include a complete history and physical examination, chest X-ray, swallowing assessment (modified barium swallow [MBS] and/or fiberoptic endoscopic evaluation of swallowing [FEES]), flexible fiberoptic laryngoscopy, and the gold standard of operative endoscopy for palpation of the interarytenoid area. Other diagnostic adjuncts may include measurement of lipid laden macrophage levels obtained by bronchial alveolar lavage[11] or interarytenoid injection.[12–15]

Treatment

The goals of management of laryngeal clefts are 2-fold—(1) to resolve the feeding impairment and (2) to minimize respiratory complications. A multidisciplinary approach that includes assessment by an otolaryngologist, pulmonologist, gastroenterologist, as well as speech–language pathologist or feeding specialist is suggested. If neurological impairments or comorbidities are suspected, evaluation by a neurologist may be useful.

Management of laryngeal clefts ranges from conservative management to surgical intervention. Conservative management includes feeding therapy, optimization of respiratory status, and antireflux medication if applicable. The decision to proceed to surgical intervention is dependent on both anatomy and function. The decision to surgically repair type I clefts is dependent on function alone. Some patients with type I laryngeal clefts will either not require intervention at all, or improve with conservative management. In our experience, approximately one-third of patients will improve with conservative management whereas two-thirds of patients with type 1 laryngeal clefts will go on to need surgical repair (Figure 2). Certain cleft types (II–IV) are repaired due to the presence of the cleft alone and known sequels from the anatomical abnormality.

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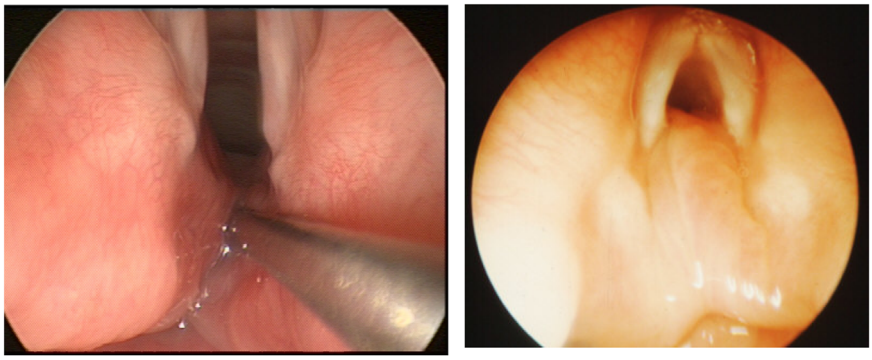


A B

Fig. 1. Endoscopy showing cleft (A) type III and type IV (B).

All patients generally require feeding therapy to address the associated swallowing impairments.[16,17] Approaches to feeding therapy may involve skill building activities such as oral-motor therapy, compensatory strategies such as the use of modified liquids or foods, modified feeding equipment such as slower or faster flow bottle nipples or open cups versus straw cups, modified feeding positioning (e.g. upright versus reclined), or modified feeding strategies (e.g. active pacing or single sips from a straw versus sequential sips). It is the role of the otolaryngologist, in consultation with the speech-language pathologist, to determine whether a patient would best be managed through feeding therapy alone or if they also require surgical repair.

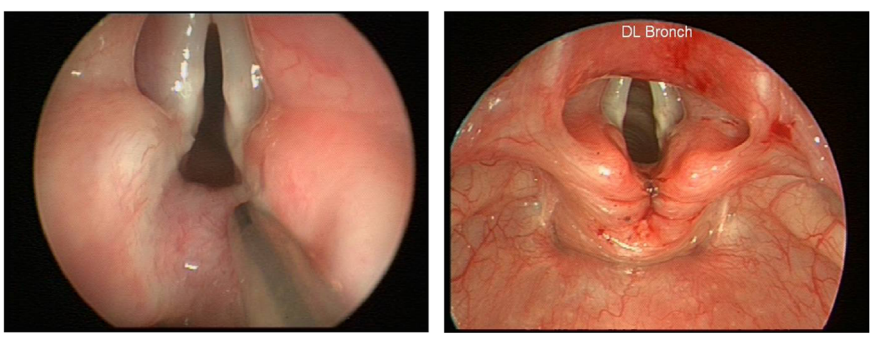
Surgical intervention may be achieved by endoscopic or open techniques. Open procedures were previously the mainstay; however, they have recently been replaced endoscopic techniques. Certain factors must be considered prior to proceeding with endoscopic repair. These include the type of cleft (types I–III are amenable depending on surgeon experience), whether adequate endoscopic visualization can be achieved due to patient anatomical factors, and the suitability of a medically-complex patient to tolerate tubeless anesthesia with spontaneous ventilation.[18] Jet ventilation or endotracheal intubation with conventional ventilation may be necessary for emergency backup ventilation.

The endoscopic repair is laser-assisted and performed with the patient in laryngeal suspension using the appropriately sized Lindholm laryngoscope with the interarytenoid space visualized in the center of the binocular microscopic field.[19] The 10-piece microlaryngeal instrument set includes 3 endoscopic needle holders (straight, left, and right), knot pusher, small and large alligator forceps, microscissors (straight and upgoing), and a vocal cord retractor. Topical 4% xylocaine is applied for topical local anesthesia, which also helps to reduce the possibility of laryngospasm during the procedure. The carbon dioxide (CO_2) laser is used to de-epithelialize the interarytenoid mucosal surface along the cleft. Appropriate laser precautions are followed. Lasering of the mucosal surface can either be done with the micromanipulator attachment for the microscope or the flexible CO_2 laser fiber (e.g., OmniGuide). Oxymetazoline on small pledgets is available to aid with hemostasis if required and to remove any excess char from the laser site. Next, vicryl sutures are placed to approximate the denuded interarytenoid mucosa. Generally, 2 vicryl sutures of 5–0 or 6–0 on a reverse cutting needle (P1) are placed for the repair of type I and II laryngeal clefts. Sutures are tied on the posterior aspect of the cleft, which avoids the placement of knots in the lumen of the airway. It is of utmost importance to completely denude the mucosa at the apex of the cleft. Failure to do so may result in a residual bridge of mucosa and resultant formation of a fistula at the apex of the repair (Figure 3). Similarly, it is also paramount to place the first suture at the most inferior extent of the cleft.

The largest series of endoscopically repaired type III laryngeal clefts was published by the senior author.[8] His series now includes 10 patients, which were all successfully repaired endoscopically. Pearls for successful surgical closure include the use of the flexible CO_2 laser fiber to ensure that the mucosal surface deep in the cleft is denuded, running the suture with several passes through the cleft, and occasionally the use of a straight Keith needle versus conventional P1 curved needle to obtain the optimal angle.

The senior author has previously utilized a transoral robotic surgical technique [20]; however, we do not currently employ this technique. Challenges of this technique include limited access to the airway when the robotic instrumentation is in place as well as lack of optimal instrumentation including suctions that adequately fit into the narrow funnel that is created by the mouth and pharynx.[21]

Open surgical approaches include the utilization of an anterior laryngofissure, lateral pharyngotomy, or thoracotomy, and may also require the use of cardiopulmonary bypass. An interposition



A B

Fig. 2. Type I cleft at diagnosis (A, pre-repair) and after repair (B, post-repair).