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Review Article

Laryngeal cleft: Evaluation and management

Douglas R. Johnston [a,*], Karen Watters [b,c], Lynne R. Ferrari [d], Reza Rahbar [b,c]

[a] Division of Otolaryngology, Nemours Alfred I. DuPont Hospital for Children, Wilmington, DE, United States
[b] Department of Pediatric Otolaryngology and Communication Enhancement, Boston Children's Hospital, Boston, MA, United States
[c] Department of Otology and Laryngology, Harvard Medical School, Boston, MA, United States
[d] Department of Anesthesiology, Boston Children's Hospital, Boston, MA, United States

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ABSTRACT

Objectives: Review the latest diagnostic and treatment modalities for laryngeal and laryngotracheoesophageal clefts as they can be a major cause of respiratory and feeding morbidity in the infant and pediatric population.
Methods: Literature review of published reports.
Results: The presentation of laryngeal cleft usually involves respiratory symptoms, such as stridor, chronic cough, aspiration, and recurrent respiratory infections. Clefts of the larynx and trachea/esophagus can occur in isolation, as part of a syndrome (Opitz-Frias, VATER/VACTERL, Pallister Hall, CHARGE), or with other associated malformations (gastrointestinal, genitourinary, cardiac, craniofacial). This publication reviews the presenting signs/symptoms, diagnostic options, prognosis, and treatment considerations based on over a decade of experience of the senior author with laryngeal clefts.
Conclusions: Type I laryngeal clefts can be managed medically or surgically depending on the degree of morbidity. Types II, III, and IV require endoscopic or open surgery to avoid chronic respiratory and feeding complications.

Contents

1. Introduction

Laryngeal and laryngotracheoesophageal clefts (hereafter referred to as laryngeal cleft) represent a spectrum of clinical symptoms and presentations based on the depth of the cleft and the overall health of the child. Type I and II laryngeal clefts often have delays in diagnosis, whereas types III and IV are likely to

Abbreviations: MBS, modified barium swallow; FEES, functional endoscopic evaluation of swallowing.

* Corresponding author at: Division of Otolaryngology, Nemours A.I. DuPont Hospital for Children, 1600 Rockland Road, Wilmington, DE 19803, United States. Tel.: +1 6107011116; fax: +1 3026515835.
E-mail address: douglas.johnston@nemours.org (D.R. Johnston).

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present within the first days of life. Likewise, a type I cleft in an otherwise healthy child may have minimal clinical impact, whereas the same cleft in a child with cardiac disease can cause significant morbidity. Therefore, clinicians who treat laryngeal clefts must treat the patient in the context of his or her overall clinical picture and recognize that laryngeal clefts represent a spectrum of disease.

Initially described by Richter in 1792, laryngeal clefts have been underdiagnosed for centuries. The first successful repair of a laryngeal cleft was performed by Petterson in 1955 [1]. A series of over 20 patients did not appear until the 1990s [2–5]. Although it has been suggested that inheritance may be autosomal dominant, it is now clear that no such associations exist [6]. A slight male preponderance is observed [2,3,5].

Estimates of the incidence of laryngeal clefts are challenging because type I clefts often go misdiagnosed for long periods and usually surface at tertiary care institutions. Nonetheless, amongst patients who have direct laryngoscopy for recurrent respiratory symptoms, the incidence ranges from 0.2 to 7.6% [3,7–12].

Embryologically, a posterior laryngeal cleft is believed to result from incomplete formation of the interarytenoid muscle with or without absence of the interarytenoid mucosa. The former is known as a submucous laryngeal cleft and the latter a type I laryngeal cleft, which does not extend beyond the level of the vocal cords. Incomplete formation of the posterior cricoid cartilage alone forms a type II cleft, and incomplete formation of the tracheoesophageal septum distal to the cricoid cartilage forms either a type III or IV cleft. Extension beyond the cricoid cartilage into the tracheoesophageal septum is a type III, and extension into the thorax is a type IV. This Benjamin and Inglis classification is the most widely used and functional of those proposed to date [13] (Fig. 1). The interarytenoid muscle (along with all intrinsic laryngeal muscles) and the cricoid cartilage are derivatives of the sixth branchial arch [14]. The above spectrum of developmental insults occurs between the fifth and sixth embryologic weeks.

Diagnosis and management of laryngeal clefts are far from uniform and can inspire controversy. Specifically, there is a lack of standardized evaluation and diagnosis; modified barium swallow (MBS) and functional endoscopic evaluation of swallowing (FEES) have an unclear role in diagnosis; the role, efficacy, and duration of feeding therapy are debated; and, lastly, the timing and type of surgical intervention is quite variable. Our diagnostic and treatment recommendations are presented herein.

2. Presentation

Presenting signs of a posterior laryngeal cleft are most often respiratory in nature. Chronic cough is common, especially with feeding, as is a variable degree of respiratory distress, depending on cleft depth. In a child with significant medical morbidities, cough may be prominent and associated with frank cyanosis. Conversely, a child with a type I cleft may have minimal disease manifestations, such as mild feeding difficulties.

In our first published series of 22 patients, roughly 90% (20/22) with type I-III clefts had respiratory symptoms, including aspiration, recurrent pneumonia, stridor, and cyanosis [4]. Our second series of 74 type I and II patients demonstrated a 50–60% rate of wheezing and chronic cough, whereas hospital respiratory admissions ranged from 15% for type I clefts to 25% for type II [5].

Other authors have reported variable rates of respiratory presenting signs in type I and II clefts, most commonly stridor, recurrent respiratory infections, and chronic cough [2,3,15]. Type III and IV clefts invariably are associated with greater respiratory symptoms with recurrent pneumonia and often excessive pulmonary mucous. They usually present within the first few days of life.

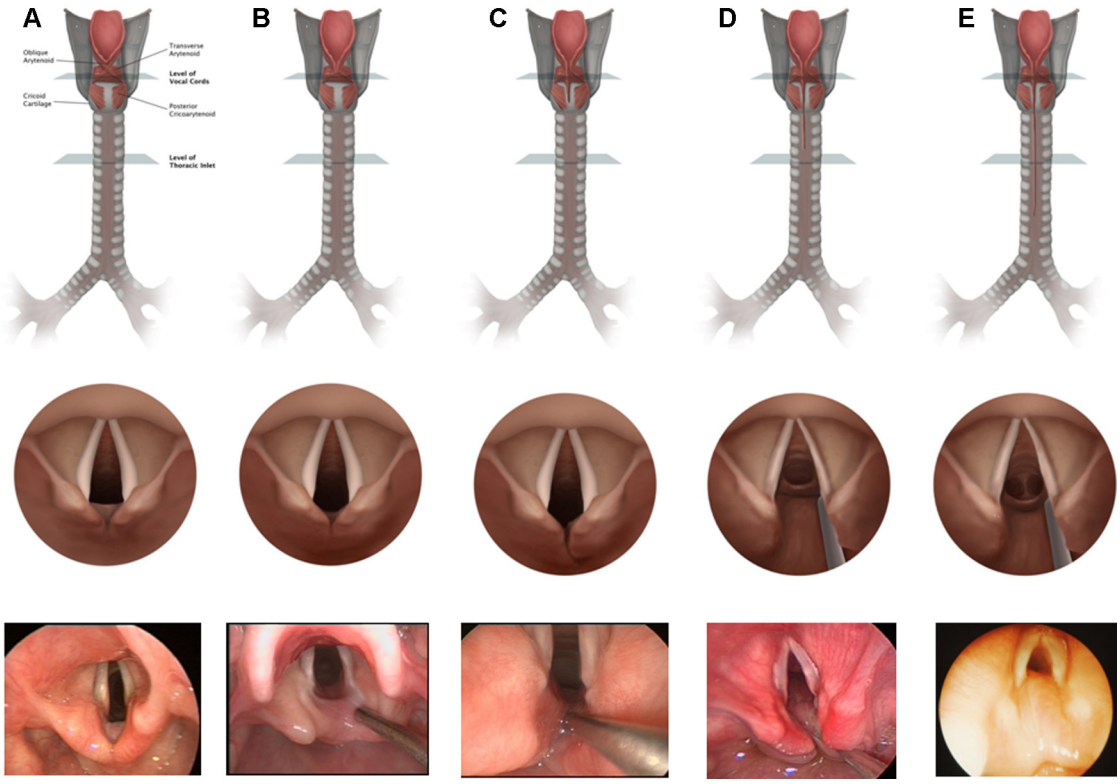

Fig. 1. Posterior view (top row), superior view (middle row), and endoscopic intraoperative view (bottom row) of Benjamin and Inglis classification of laryngeal and laryngotracheoesophageal clefts: column A, normal larynx; column B, type I extends to the level of vocal cords; column C, type II extends below vocal cords into cricoid cartilage; column D, type III extends through cricoid cartilage to cervical trachea/esophagus; column E, type IV extends to level of thoracic trachea/esophagus. *Note*: it is difficult in endoscopic view to show depth of type IV cleft as there frequently is redundant mucosa [13].