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# Type 1 Laryngeal Clefts: An Updated Review

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**Abstract** Difficulty with feeding is not an uncommon presenting symptom in patients seeking pediatric otolaryngology consultation, and the differential diagnosis is lengthy. While severe laryngeal clefts often present close to the time of birth, type 1 clefts may present after several years of life. A standardized approach to the patient with a type 1 laryngeal cleft (T1LC) has not yet been well described. A debate still exists regarding the clinical significance of a T1LC as well as the ideal management.

**Keywords** Laryngeal cleft · Aspiration · Chronic cough · Airway anomaly · Congenital airway anomaly

## Introduction

### Background

The laryngeal complex is formed from the 4th and 6th pharyngeal arches. During developmental weeks 5–6, the laryngotracheal groove and arytenoid swellings form. These structures further develop into the larynx as the trachea and esophagus differentiate. During this time, the tracheoesophageal septum develops, separating the airway from the hypopharynx and esophagus. Failure of this septum to completely form may lead to abnormalities including a laryngeal cleft or tracheoesophageal fistula [1–3] Laryngeal clefts were first described by Richter in 1792, and occur in 1:10,000–1:20,000 live births with a slight male predominance [3, 4, 5••, 6, 7, 8•, 9, 10].

The Benjamin-Inglis classification system is the most widely accepted and describes four types of LCs. A type 1 cleft is described as a deep notch which extends to, but not through the level of the vocal cords. A type 2 cleft extends into the cricoid cartilage while a type 3 extends through the entire cricoid, with or without extension into the posterior tracheal wall, but not beyond the thoracic inlet. A type 4 cleft extends through the posterior tracheal wall and beyond the thoracic inlet [4, 5••, 6, 8•, 9, 11••] A T1LC is the most common form, and has been reported to be present in up to 7.6 % of patients undergoing operative airway evaluation for concerns of aspiration [4, 11••]. T1LC presents a more challenging diagnostic and treatment dilemma due to the inability to identify them on flexible laryngoscopy, intermittent aspiration which may not be noted by MBS or FEES, and variable response to management.

### Presentation

Children with a T1LC may present as early as within the first month of life to teenage years, but most frequently are seen by an otolaryngologist around age 1–3 years of age [4, 5••, 6, 11••, 12] Difficulty with feeding is the most common presenting symptom in patients with a LC1. This may include frank aspiration of thin liquids, coughing/choking with feeds or cyanotic spells while feeding. Other common presenting symptoms include chronic cough, recurrent respiratory infections, hoarseness, nocturnal cough, stridor, or wheezing [3, 7, 11••, 12]. Although incidentally noted

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**Table 1** Most common syndromes associated with T1LC

| Syndrome | Features |
|---|---|
| CHARGE | Coloboma of eyes, heart abnormalities, choanal atresia, growth and mental retardation, urogenital and ear abnormalities |
| VACTERL | Vertebral abnormalities, anal atresia, cardiac abnormalities, tracheoesophageal fistula, ear, renal and limb abnormalities |
| Opitz frias | Cleft lip and palate, hypertelorism, hypospadias |
| Pallister hall | Hypothalamus/pituitary abnormalities, poly- and syn-dactyly, imperforate anus, renal, limb and ear abnormalities |
| PHACES syndrome | Posterior fossa malformations, hemangiomas, arterial anomalies, cardiac defects, eye abnormalities, sternal clefting |
| Trisomy 21 | Developmental delay, craniofacial and endocrine abnormalities, macroglossa, strabismus, congenital cardiac defects |

T1LC's have been reported, the true incidence of asymptomatic disease is difficult to attain because most children undergoing operative evaluation have some degree of a respiratory or swallowing complaints [9].

Approximately 35–50 % of children with a laryngeal cleft have an associated congenital anomaly or syndrome, and up to 90 % carry concomitant diagnoses such as prematurity, neurodevelopmental delay, laryngomalacia, gastroesophageal reflux disease (GERD), or reactive airway disease [3, 4, 6, 8•, 9, 11••, 12] (Table 1). Due to the wide variety of associated pathologies seen in patients and the irregularity which it appears to be categorized, the literature varies in the prognostic value of such abnormalities seen in patients with T1LC. Several authors have assessed outcomes relative to medical and/or congenital comorbidities without identifying concurrent diagnoses that carry a prognostic value [3, 5••, 6, 7]

However, Ojha et al. [11••] studied the outcomes of 42 patients with a T1LC and separated the patients with comorbidities that put them at risk of aspiration from those with comorbidities unrelated to aspiration. Patients were managed either conservatively with a thickened diet or with endoscopic repair. All of the children who were otherwise healthy (12 %) were managed with conservative care alone. Twenty-seven children (64 %) required surgical management, and 78 % of these patients were ultimately advanced to a regular diet. Six patients failed surgical treatment; all of these had comorbidities directly relating to an increased risk of aspiration [11••].

## Diagnostic Testing

Because a T1LC can present with a wide variety of symptoms, a high index of suspicion must be present in order to prevent a delay in diagnosis and management. Microlaryngoscopy with palpation of the posterior interarytenoid space is considered the gold standard for diagnosis (Fig. 1a, b). However, flexible fiberoptic laryngoscopy should be performed as well in order to diagnose concurrent dynamic pathologies such as laryngomalacia or vocal cord immobility.

Either a functional endoscopic evaluation of swallowing (FEES) or modified barium swallow (MBS) should be performed as part of the initial assessment in any child with significant dysphagia. Evaluation by a speech pathologist has both diagnostic and therapeutic benefit as well. Although many patients with a T1LC aspirate only intermittently, a swallowing evaluation is important to assess overall functional swallowing and identify any concurrent deficits. The speech pathologist should evaluate swallowing with various consistencies and behavioral/positional modifications during these studies. Both exams can assess laryngeal penetration and aspiration, however, only the MBS can evaluate the oral, pharyngeal, and esophageal phases of swallowing. However, FEES provides many benefits as well (such as a detailed anatomic exam, evaluation of ability to handle secretions, and direct sensation testing).

Both FEES and MBS can serve as important tools for more objective pre- and post-operative data, to follow patients who are treated conservatively, and to formalize research methodology. Horn et al. [5••] proposed a point scale using MBS allowing them to better score the patient's level of aspiration. Three consistencies were graded; thin, nectar, and honey thick. Patients were assigned a score of 0–3 (normal, laryngeal penetration, microaspiration, or frank aspiration) for each consistency. Patients with a normal swallow in all consistencies were given a score of 0 and those who experienced frank aspiration in all consistencies were given a score of 9. Although this instrument requires validation, it could be modified for FEES and potentially be used as a standardized way of quantifying aspiration. Further evaluation of its clinical correlation to aspiration may be an area of research interest.

The lipid laden macrophage index (LLMI) is another area of research in aspiration and chronic aerodigestive inflammation [14, 15]. LLMI measures the accumulation of lipids within the alveolar macrophages, indicating the presence of inflammation. Kieran et al. [14] studied the LLMI in patients with a type 1 and type 2 laryngeal cleft. The group performed flexible bronchoscopy with bronchoalveolar lavage in 44 patients with laryngeal clefts (31 type 1 and 13 type 2). They found a significantly higher LLMI in patients with a type 2 cleft than a type 1 (81.8 vs. 44.9) and in patients with a history of reactive airway disease (84.6 with RAD vs. 48.5 without). No correlation was found between the LLMI and presence of aspiration on

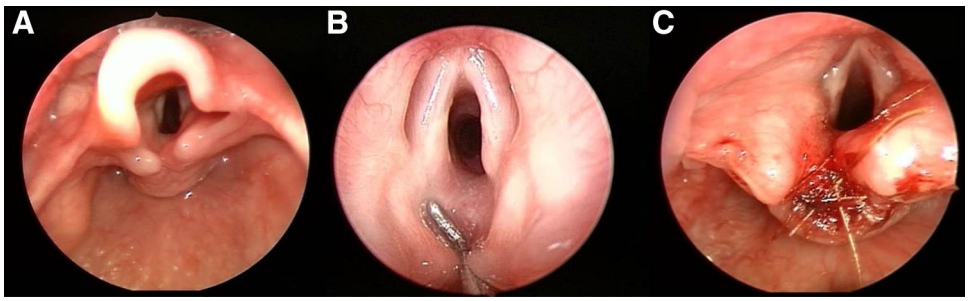


**Fig. 1** **a–c** Inability to visualize the laryngeal cleft without probing (**a**) and identification of the T1LC with palpation of the interarytenoid space with a right angle probe (**b**). Immediately following repair of a T1LC using the microflap technique (**c**)

MBS, history of GERD, chronic cough, or pneumonia. Although there is a paucity of normative data, the authors propose that the LLMI may be used as part of an algorithm to determine severity of disease and urgency to surgically repair the cleft.

The presence of a laryngeal cleft must be on the differential when evaluating a patient with chronic or recurrent difficulties with feeding or respiration. A thorough history and physical exam including flexible laryngoscopy, MBS or FEES, and operative airway exam are required to diagnose these patients and optimally manage their symptoms.

## Management

Management is the most debated topic involving T1LC's. Several authors have proposed diagnostic and management algorithms, but a united consensus, especially regarding the timing and type of repair, has yet to be determined [3, 4, 6, 11••].

### Conservative Management

The first line therapy for patients who have a T1LC is conservative management. This includes anti-reflux medication as well as speech therapy with thickening of feeds, behavioral and positional modifications. Additionally, comorbid conditions such as reactive airway disease should be optimized. Approximately 35–55 % of patients improve over time with conservative measures; however, this number ranges from 0 to 100 % in the literature proving the difficulty in establishing guidelines for management [6, 8•]. Children experiencing chronic symptoms in spite of conservative management should be considered as candidates for surgical management.

### Endoscopic Repair

Definitive surgical repair of a T1LC consists of endoscopic closure of the cleft using either a microflap technique or CO2 laser. The operative microscope with laryngeal suspension and a vocal cord spreader is used to obtain optimal visualization of the posterior glottis. If possible, repair is done under spontaneous ventilation without the need for intubation [16]. The microflap technique is performed first with a U-shaped incision in the cleft and undermining of the mucosa. Two posterior and anterior flaps are created and then sewn together in two layers (Fig. 1c) [7].

With the CO2 laser technique, the interarytenoid mucosa is thoroughly denuded and any char removed. The cleft is then closed with a single layer of deep absorbable sutures and the knot is directed posteriorly. This is thought to prevent granuloma formation within the glottis [6, 7, 17].

More recently, the use of transoral robotic surgery (TORS) has been implemented in the endoscopic repair of laryngeal clefts [18, 19] The largest study, published in 2014, reported the feasibility of TORS in 5 patients. All patients in the group underwent oral or nasal tracheal intubation and exposure was obtained with the Feyh–Kastenbauer or Dingman retractor. The mucosa was denuded with cautery and the cleft reapproximated with absorbable suture. Total operative time ranged from 173 min for their first case to 105 min for the fifth case. By 4 weeks of time in follow-up, all 5 patients tolerated all consistencies of liquids [18]. As TORS has become a more widely used technique in otolaryngology, further cost and resource analysis must be performed to understand its optimal applications.

Regardless of technique, patients undergoing endoscopic repair should be maintained on an anti-reflux regimen immediately after surgery. There does not appear to be a consensus regarding post-operative intubation, as some authors describe outpatient surgery, while others will keep the child intubated for 48–72 h. This is likely best to assess on a case by case basis.

Successful endoscopic repair has been quoted as high as a 94 %, but most reports range from 50 to 75 % [4]. Interestingly, several authors report a delay in symptomatic improvement [7, 11••]. Ohja et al. [11••] studied 42 T1LC patients who failed conservative therapy and underwent endoscopic repair. Post-operative MBS was performed at

6 weeks and patients were examined clinically at 6 weeks, 3, 6, and 12 months. At the 6 week follow-up, 70 % of patients were improving but none had complete resolution of their symptoms. At 3, 6, and 12 month follow-up visits 26, 37, and 44 % of patients demonstrated resolution of their symptoms, respectively. In total, 78 % of patients experienced improvement or resolution at the 12 month follow-up. In many cases, aspiration may be multifactorial with repair allowing for a slow compensation over time.

## Injection Laryngoplasty

Predicting which patients will have a successful endoscopic repair continues to be a challenge. And, because of this dilemma, a "diagnostic" injection laryngoplasty (IL) was proposed in 2000 by Kennedy and colleagues [20]. They injected gelfoam into the posterior supraglottis of 8 patients with T1LC and found all patients to have an initial improvement in their symptoms. One patient who required multiple injections due to a recurrence of symptoms ultimately underwent a successful endoscopic repair. Since that time, however, larger studies with longer follow-up have been published and a success rate of 100 % has not been replicated.

In 2011, Cohen et al. [3] reviewed 16 patients who underwent IL with either sodium carboxymethylcellulose aqueous gel or gelfoam. Nine patients (56 %) experienced complete resolution of their symptoms and were noted to have a normal post-operative MBS. Of the remaining 44 %, 4 patients had some improvement and 3 were considered failures. Five of the 9 patients who initially underwent IL had recurrence of symptoms after about 3.5 months, and went on to have successful endoscopic repair. In 2012, Mangat et al. [8•] studied 18 children who underwent IL with gelfoam or hyaluronic acid-based fillers, only two of whom had significant medical comorbidities (fetal alcohol syndrome and intrauterine drug exposure). This group had a mean follow-up of 17 months and reported one complication of post-

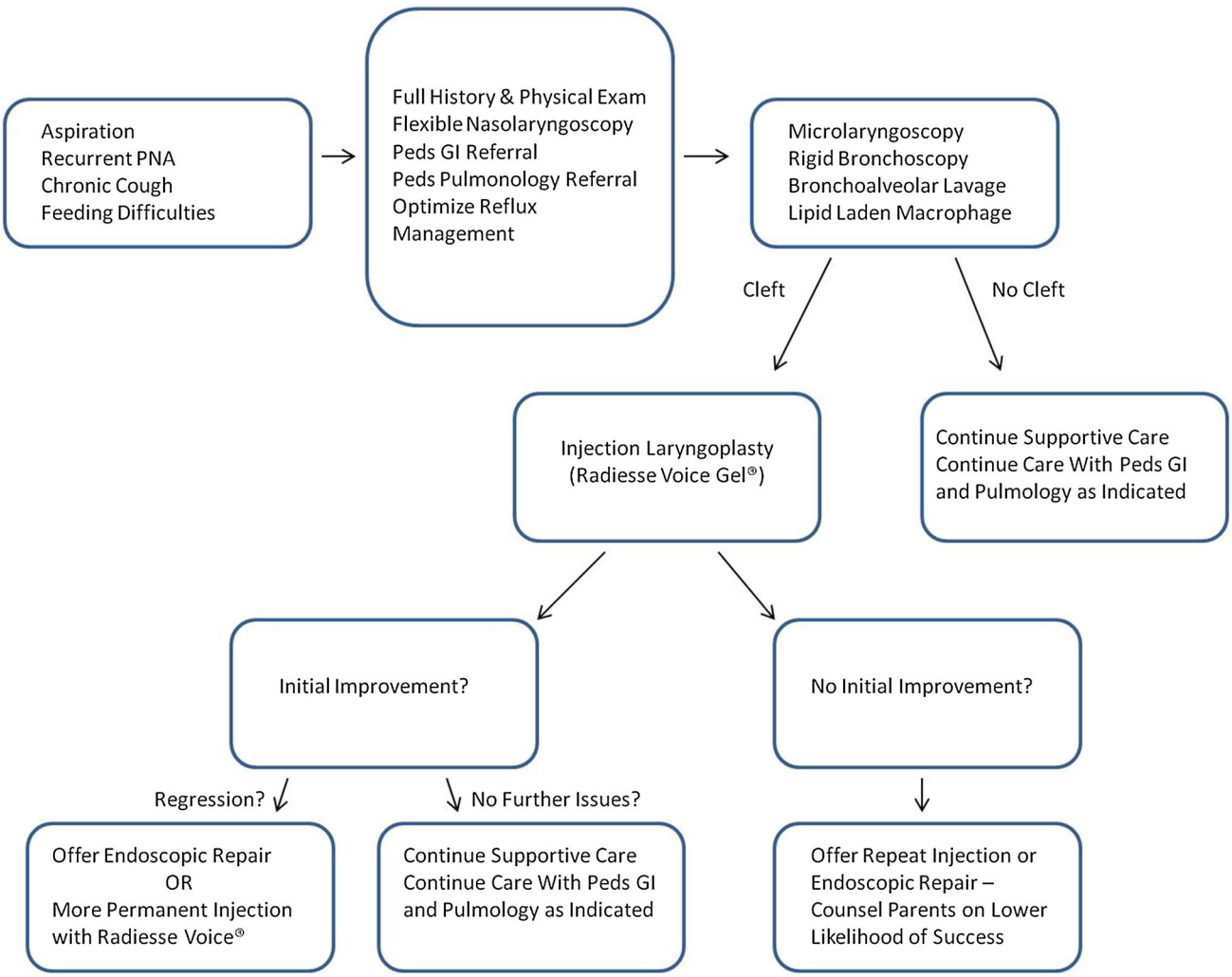


**Fig. 2** Algorithm for type 1 laryngeal cleft management

operative swelling requiring hospital admission and systemic steroids. Thirteen (72 %) of patients experienced improvement in swallowing dysfunction based on clinical exam, parent report, and/or MBS. Seven patients underwent repeat injections and only one ultimately required endoscopic repair. The authors recognize they selected healthier patients without significant comorbidities, but do not describe proposed reasons for the 5 patients who had no improvement.

Most recently, Horn et al. [5••] proposed IL for children suffering from chronic aspiration with an unknown etiology. Parents were offered diagnostic microlaryngoscopy for their child, and an IL was performed at the time of surgery regardless of the presence or absence of a laryngeal cleft. An MBS was performed on all children prior to intervention and within 2 weeks post-operatively. The MBS was scored as discussed above. Thirty patients underwent IL with sodium carboxymethylcellulose gel for chronic aspiration and 28 with appropriate follow-up were analyzed. Children were considered "improved" if they could safely swallow thin liquids. 57.1 % of patients experienced improvement, but interestingly only 31.3 % of these patients had a laryngeal cleft. Thus, 50 % of patients without a laryngeal cleft and chronic aspiration, improved with IL. The authors argue that some patients may have a submucous or physiologic cleft with an incompetent interarytenoid larynx, and injection followed by resorption allows for patient to be "weaned" from the augmentation [5••]. Both horizontal and vertical "occult" clefts with intact mucosa have been reported in post mortem exams; clearly these would be very difficult to diagnose at the time of an operative airway exam [21]. Although the presence of a cleft appeared to be a prognostic indicator (33.1 vs. 8.3 % of patients who improved had a T1LC), it was not statistically significant. Additionally, 5 of 6 T1LC patients versus 11 of 22 non-T1LC patients improved after IL, but again, this did not reach statistical significance. Not surprisingly, mean MBS scores were significantly lower in the patients who improved vs those who did not, which aids in validating the use of their scoring methods. Five total endoscopic repairs were done in this group, and only children who initially responded to IL had resolution of their symptoms supporting the use of IL as both a treatment and prognostic modality [5••].

As the predictability of surgical responders remains a challenge, IL has become part of the treatment algorithm at our institution (Fig. 2).

## Conclusion

As pediatric otolaryngologists have become more aware of laryngeal clefts, operative evaluation in the patient with chronic feeding difficulties has become routine. The timing of the exam relative to a formal swallow evaluation, and evaluation by our pediatric gastroenterology and pulmonology colleagues has not been standardized. The use of IL as both a diagnostic and therapeutic tool is still a relatively new concept and warrants further investigation with regard to its timing, ideal materials, and prognostic abilities. It appears that the children with fewer comorbidities are more likely to have success with conservative management. However, the children with more severe comorbidities are the patients most likely to fail endoscopic surgical management. Therefore, this group of patients may currently require the most attention in future research studies.

### Compliance with Ethics Guidelines

**Conflict of Interest** Karen Hawley and David White declare that they have no conflict of interest.

**Human and Animal Rights and Informed Consent** This article does not contain any studies with animal subjects performed by any of the authors. With regard to the authors' research cited in this paper, all procedures were followed in accordance with the ethical standards of the responsible committee on human experimentation and with the Helsinki Declaration of 1975, as revised in 2000 and 2008. IRB approval is noted in all of the authors' cited works.

## References

Papers of particular interest, published recently, have been highlighted as:
- • Of importance
- •• Of major importance


1. Cochard LR. Netter's atlas of human embryology. Updated Ed. Chapter 5. The respiratory system. Chicago: Saunders Elsevier; 2012. p. 113–129.
2. Cochard LR. Netter's atlas of human embryology. Updated Ed. Chapter 6. The gastrointestinal system and abdominal wall. Chicago: Saunders Elsevier; 2012. p. 131–156.
3. Cohen MS, Zhuang L, Simons JP, Chi DH, Maguire RC, Mehta DK. Injection laryngoplasty for type 1 laryngeal cleft in children. Otolaryngol Head Neck Surg. 2011;144(5):789–93.
4. Chien W, Ashland J, Haver K, Hardy SC, Curren P, Hartnick CJ. Type 1 laryngeal cleft: establishing a functional and management algorithm. IJPO. 2006;70:2073–9.
5. •• Horn DL, DeMarre K, Parikh SR. Interarytenoid sodium carboxymehtylcellulose gel injection for management of pediatric aspiration. Ann Otol Rhinol Laryngol. 2014;123:852–8. *Novel approach to pediatric aspiration.*
6. Johnston DR, Watters K, Ferrari LR, Rahbar R. Laryngeal cleft: evaluation and management. IJPO. 2014;78:905–11.
7. Ketcham AS, Smith JE, Lee FS, Halstead LA, White DR. Clinical course following endoscopic repair of type 1 laryngeal clefts. IJPO. 2008;72:1261–7.
8. • Mangat HS, El-Hakim H. Injection augmentation of type 1 laryngeal clefts. Otolaryngol Head Neck Surg. 2012;146(5):764–8. *Largest injection laryngoplasty study for laryngeal cleft patients to date.*

9. Parsons DS, Stivers FE, Giovenetto DR, Phillips SE. Type I posterior laryngeal clefts. Laryngoscope. 1998;108:403–10.
10. Watters K, Russell J. Diagnosis and management of type 1 laryngeal cleft. IJPO. 2003;76:591–6.
11. •• Ojha S, Ashland JE, Hersh C, Ramarishna J, Maurer R, Hartnick CJ. Type 1 laryngeal cleft—a multidimensional management algorithm. JAMA Otolaryngol Head Neck Surg. 2014;140(1):34–40. *Excellent study design assessing timing of improvement and specific comorbidities*.
12. Rahbar R, Chen JL, Rosen RL, Lowry KC, Simon DM, Perez JA, Buonomo C, Ferrari LR, Katz ES. Endoscopic repair of laryngeal cleft type I and type II: when and why? Laryngoscope. 2009;119:1797–802.
13. Brady S, Donzelli J. The modified barium swallow and the functional endoscopic evaluation of swallowing. Otolaryngol Clin N Am. 2013;46:1009–22.
14. Kieran SM, Katz E, Rosen R, Khatwa U, Martin T, Rahbar R. The lipid laden macrophage index as a marker of aspiration in patients with type I and II laryngeal clefts. IJPO. 2010;74:743–6.
15. Reilly BK, Katz ES, Misono AS, Khatwa U, Didas A, Huang L, Haver K, Rahbar R. Utilization of liped-laden macrophage index in evaluation of aerodigestive disorders. Laryngoscope. 2011;121(5):1055–9.
16. Ferrari L, Zurakowski D, Solari J, Rahbar R. Anesthesia for laryngeal cleft surgery. Ped Anesth. 2013;23:334–41.
17. Watters K, Ferrari L, Rahbar R. Minimally invasive approach to laryngeal cleft. Laryngoscope. 2013;123:264–8.
18. Ferrell JK, Roy S, Karni RJ, Yuksel S. Applications for transoral robotic surgery in the pediatric airway. Laryngoscope. 2014;124:2630–5.
19. Leonardis RL, Duvvuri U, Mehta D. Transoral robotic-assisted laryngeal cleft repair in the pediatric patient. Laryngoscope. 2014;124:2167–9.
20. Kennedy CA, Heimbach M, Rimell FL. Diagnosis and determination of the clinical significance of type IA laryngeal clefts by gelfoam injection. Ann Otol Rhinol Laryngol. 2000;109(11):991–5.
21. Tucker GF, Maddalozzo J. "Occult" posterior laryngeal cleft. Laryngoscope. 1987;97(6):701–4.