Type I-II Laryngeal Cleft: Clinical Course and Outcome

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ABSTRACT: **Background:** Laryngeal cleft (LC) is a rare congenital anomaly manifesting in a variety of symptoms, including swallowing disorders and aspirations, dyspnea, stridor and hoarseness. The mild forms (types I-II) may be underdiagnosed, leading to protracted symptomatology and morbidity.

Objectives: To evaluate the diagnostic process, clinical course, management and outcome in children with type I-II laryngeal clefts.

Methods: We conducted a retrospective case analysis for the years 2005–2012 in a tertiary referral center.

Results: Seven children were reviewed: five boys and two girls ranging in age from birth to 5 years. The most common presenting symptoms were cough, aspirations and pneumonia. Evaluation procedures included fiber-optic laryngoscopy (FOL), direct laryngoscopy (DL) and videofluoroscopy. Other pathologies were seen in three children. Six children underwent successful endoscopic surgery and one child was treated conservatively. The postoperative clinical course was uneventful in most of the cases.

Conclusions: Types I-II LC should be considered in the differential diagnosis of children presenting with protracted cough and aspirations. DL is crucial for establishing the diagnosis. Endoscopic surgery is safe and should be applied promptly when conservative measures fail.

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KEY WORDS: laryngeal cleft (LC), fiber-optic laryngoscopy (FOL), stridor, aspirations, direct laryngoscopy (DL)

Laryngeal cleft (LC) constitutes less than 0.3% of all congenital laryngeal anomalies (approximately 1:2000 live births) [1,2]. LC is the result of failed fusion of the tracheo-esophageal folds or the dorsum of the cricoid ring during gestational weeks 5–7 [3,4]. Benjamin and Inglis [5] divided LC into four subtypes based on the caudal extension from the interarytenoid area towards the carina. Type I LC is considered supraglottic and involves the interarytenoid region only, while type II involves the cricoid lamina. Types III and IV extend through the cricoid lamina down to the cervical and intrathoracic trachea, respectively [5].

Various congenital malformations, syndromic and non-syndromic, have been reported in association with LC. These include gastrointestinal defects such as tracheo-esophageal fistula (TEF), cardiac anomalies, and anomalies of the great vessels and genitourinary system [1,6].

Symptoms are non-specific, varying from mild stridor to massive aspirations with recurrent episodes of pneumonia, depending on the severity of the cleft [7]. Thus, the key to early diagnosis of mild cases is a high index of suspicion and thorough laryngeal evaluation [4]. In many type I LC patients with only minor symptoms, conservative measures, based on a proper feeding regimen including food consistency, might be sufficient [8]. However, when symptomatic, closure of the cleft is mandatory.

This report presents our experience with types I-II LC, emphasizing the apparently indolent and obscure course of the disease and its possible management, with the aim of raising awareness regarding LC in the differential diagnosis of protracted airway disease.

PATIENTS AND METHODS

Seven children were diagnosed with LC between the years 2005 and 2012. Three had type I and four had type II. The mean age at diagnosis of the five boys and two girls was 15 months (range 0–48 months) and mean age at surgery 24 months (range 3–49 months). Patients' gender, cleft type, associated pathologies, age at diagnosis, age at surgery and diagnostic procedures are summarized in Table 1.

DIAGNOSTIC PROCEDURES

Fiber-optic laryngoscopy (FOL) followed by direct laryngoscopy (DL) were performed in all children prior to surgery. Direct laryngoscopy enables palpation and verification of the depth of the lesion. Two children were evaluated by DL alone and one child underwent videofluoroscopy [Table 1].

OPERATIVE TECHNIQUE

Endoscopic surgery was performed via DL. The child was prepared in a supine position with mild head extension. A rigid Lindholm laryngoscope was used to expose the posterior laryngeal wall with proximal jet ventilation (one patient was ventilated through a tracheotomy). Marcaine with adrenaline (1:100,000) was injected into the mucosa overlying the cleft. Cold instruments were used for verification of the cleft's depth, making mucosal incisions along the cleft down to its apex, and

Table 1. Types I and II laryngeal cleft: patient characteristics

Patient	Gender	Cleft type	Associated pathologies	Age at diagnosis (months)	Age at surgery (months)	Diagnostic procedures
1	M	II		Birth	3	DL, video-fluoroscopy
2	M	II		6	18	FOL, DL
3	M	II	TEF, supraglottic collapse, tracheotomy	18	24	FOL, DL
4	M	II		36	60	FOL, DL
5	F	I	VACTERL association	Birth	4	FOL
6	F	I	IUGR, prematurity, PIE, BPD, PDA, developmental delay	16	–	FOL
7	M	I		48	49	DL

DL = direct laryngoscopy, FOL = fiber-optic laryngoscopy, TEF = tracheo-esophageal fistula, IUGR = intrauterine growth restriction, PIE = pulmonary interstitial emphysema, BPD = bronchopulmonary dysplasia, PDA = patent ductus arteriosus

Table 2. Types I and II laryngeal cleft: presenting symptoms

Patient	Cleft type	Stridor	Dysphonia	Aspirations	Cough	Pneumonia
1	II	-	-	+	+	+
2	II	-	-	+	+	+
3	II	-	+	+	+	+
4	II	-	-	-	+	+
5	I	+	-	+	+	-
6	I	-	+	+	+	+
7	I	+	+	+	-	+

+ = present, - = absent

raising anterior and posterior mucosal flaps. The mucosal layers (cleft 'shoulders') were apposed (the anterior-esophageal layer first) with interrupted 5-0 polyglactin absorbable sutures. Liquid oral intake was initiated on postoperative days 1–2. Postoperative H2 blockers were used routinely. Patients were monitored postoperatively in a pediatric intensive care unit (PICU) and later in a pediatric ward.

RESULTS

LC presenting symptoms consisted of airway and feeding difficulties (aspirations and cough), leading to recurrent pulmonary infections in the majority of patients. Stridor and dysphonia were less common [Table 2]. All but one patient, who already presented with a tracheotomy, were operated with proximal jet ventilation. Liquid oral intake was resumed on postoperative days 1–2. Mean PICU stay was 1 day.

Postoperative complications were considered minor, including fever of 48 hours in one child, assuming an episode of aspiration pneumonia; and another child required intubation with assisted ventilation for 24 hours. A 6 month follow-up was uneventful and all children resumed oral feeding. Three children had the following associated anomalies:

Child # 3 underwent tracheotomy due to recurrent episodes of aspiration pneumonia. He underwent repair of TEF as a neonate and type II LC was corrected successfully at age 2 years. Decanulation was still delayed due to laryngomalacia with supraglottic collapse. CO_2 laser supraglottoplasty was performed 8 months following the LC correction, with successful decanulation 3 months later.

Child # 5 was diagnosed at birth with VACTERL association and underwent TEF repair and colostomy due to imperforated anus. In addition, she presented with biphasic stridor mainly while crying, and continuous aspirations. FOL revealed type I LC that was operated successfully at age 4 months. She was kept intubated for 1 postoperative day. Occasional episodes of aspirations still continued during feeding for another 3 months. One year of follow-up was uneventful.

Child # 6 was diagnosed with severe intrauterine growth retardation, born prematurely in the 28th week weighing 740 g. She was alternately intubated for a total of 40 days due to acidosis, pulmonary interstitial emphysema and bronchopulmonary dysplasia. Patent ductus arteriosus was corrected in the 2nd week of age. In addition, she presented with feeding difficulties which required total peripheral nutrition until age 2 months. Her medical history also included three episodes of pneumonia that were considered unrelated to aspirations. At age 16 months, a type I LC was detected and was managed conservatively by means of a thickened feeding regimen and careful swallowing. By age 3 years, her feeding difficulties had resolved completely and the only remaining symptom was mild hoarseness.

DISCUSSION

Laryngeal cleft is a rare congenital disorder that may be overlooked by the inexperienced physician [9]. The most common presenting symptoms in our series were cough, recurrent aspirations and pneumonia. Yet, the variety of presentations and their severity differ significantly between reports: Glossop et al. [10] claimed that aspirations may be absent or of minor severity in grade I clefts, while Parsons et al. [11] emphasized symptoms related to laryngeal incompetence including micro-aspirations, especially of thin liquids, with protracted cough mimicking asthma-like symptoms, recurrent lower respiratory tract infections, and choking during feeding; yet, all these symptoms were often subtle. In another report, Evans and co-authors [12] observed that cyanotic attacks with feeding were the most common presenting symptom in type I clefts (48%) followed

by inspiratory stridor (44%) and recurrent chest infections (24%). Varying degrees of dysphonia (including a husky voice or a weak abnormal cry and voiceless crying) were also noted. Van der Doef et al. [7] reviewed the literature on type I LC and advocated considering this pathology in children with congenital stridor and laryngomalacia, particularly when stridor is accompanied by feeding difficulties. As previously reported, the most common presenting symptoms are non-specific and demand a high level of suspicion in protracted cases.

The majority of patients can be diagnosed by FOL in an outpatient setting, or, at least, referred for further evaluation in light of ill-defined laryngeal findings. In our experience, definitive diagnosis should be established by DL, enabling unlimited exposure and palpation of the inter-arytenoid area. Nevertheless, a delay in diagnosis still occurs, ranging from weeks to years [13], inflicting chronic cough and feeding problems, aspirations with recurrent episodes of pneumonia, recurrent hospitalizations and failure to thrive. It is reasonable to assume that the association of other pathologies may mask small LC with minor symptoms.

Successful conservative treatment, based on proper positioning during feeding and thickened feeding regimen, was previously described in type I LC [11,12]. One of our patients was successfully treated accordingly. All other patients underwent uneventful and successful surgical interventions. We assume that a delay in diagnosis (found in some of our patients) rendered them already resistant to conservative treatment trials.

The majority of types I-II LC can be managed endoscopically. Recently, Sandu and Monnier [14] reported a series of four children with type III LC operated endoscopically without a tracheotomy. Still, advanced type II and deeper clefts may demand a combined cervical (laryngofissure or lateral pharyngotomy) and thoracic approach with tissue grafts interposition [6,13].

Mucosal incisions are performed by either cold instrumentation or CO_2 laser. However, it is important to completely remove the mucosa at the apex of the cleft to prevent a residual mucosal bridge that would lead to a persistent fistula at the bottom of the cleft [13].

The use of jet ventilation enabled optimal exposure and access to the posterior laryngeal wall. This was very useful in manipulating and apposing the mucosal flaps in a manner that restored near normal anatomy and function with minimal trauma. Liquid oral intake was regained on the first or second postoperative days. Only one patient needed reintubation and there were no long-term sequelae.

Follow-up periodic airway evaluation and repeated swallowing studies are quite limited among infants in our outpatient setting. It is reasonable to assume that resuming normal feeding without coughing or aspirations indicates adequate airway protection.

CONCLUSIONS

Laryngeal cleft is a rare congenital malformation, manifesting as non-specific respiratory and feeding difficulties that may not respond to conservative management. Early diagnosis relies on a high index of suspicion, while definitive diagnosis should be made by direct laryngoscopy. Endoscopic correction of type I-II LC should be considered simple and safe and should be promptly applied when conservative measures fail.

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References

1. Eriksen C, Zwillenberg D, Robinson N. Diagnosis and management of cleft larynx. Literature review and case report. *Ann Otol Rhinol Laryngol* 1990; 99 (9 Pt 1): 703-8.
2. DuBois JJ, Pokorny WJ, Harberg FJ, Smith RJ. Current management of laryngeal and laryngotracheoesophageal clefts. *J Pediatr Surg* 1990; 25 (8): 855-60.
3. Blumberg JB, Stevenson JK, Lemire RJ, Boyden EA. Laryngotracheoesophageal cleft, the embryologic implications: review of the literature. *Surgery* 1965; 57: 559-66.
4. Carr MM, Clarke KD, Webber E, Giacomantonio M. Congenital laryngotracheoesophageal cleft. *J Otolaryngol* 1999; 28 (2): 112-17.
5. Benjamin B, Inglis A. Minor congenital laryngeal clefts: diagnosis and classification. *Ann Otol Rhinol Laryngol* 1989; 98 (6): 417-20.
6. Kubba H, Gibson D, Bailey M, Hartley B. Techniques and outcomes of laryngeal cleft repair: an update to the Great Ormond Street Hospital series. *Ann Otol Rhinol Laryngol* 2005; 114 (4): 309-13.
7. van der Doef HP, Yntema JB, van den Hoogen FJ, Marres HA. Clinical aspects of type 1 posterior laryngeal clefts: literature review and a report of 31 patients. *Laryngoscope* 2007; 117 (5): 859-63.
8. Chien W, Ashland J, Haver K, Hardy SC, Curren P, Hartnick CJ. Type 1 laryngeal cleft: establishing a functional diagnostic and management algorithm. *Int J Pediatr Otorhinolaryngol* 2006; 70 (12): 2073-9.
9. Watters K, Russell J. Diagnosis and management of type 1 laryngeal cleft. Int *J Pediatr Otorhinolaryngol* 2003; 67 (6): 591-6.
10. Glossop LP, Smith RJ, Evans JN. Posterior laryngeal cleft: an analysis of ten cases. *Int J Pediatr Otorhinolaryngol* 1984; 7 (2): 133-43.
11. Parsons DS, Stivers FE, Giovanetto DR, Phillips SE. Type I posterior laryngeal clefts. *Laryngoscope* 1998; 108 (3): 403-10.
12. Evans KL, Courteney-Harris R, Bailey CM, Evans JN, Parsons DS. Management of posterior laryngeal and laryngotracheoesophageal clefts. *Arch Otolaryngol Head Neck Surg* 1995; 121 (12): 1380-5.
13. Rahbar R, Rouillon I, Roger G, et al. The presentation and management of laryngeal cleft: a 10-year experience. *Arch Otolaryngol Head Neck Surg* 2006; 132 (12): 1335-41.
14. Sandu K, Monnier P. Endoscopic laryngotracheal cleft repair without tracheotomy or intubation. *Laryngoscope* 2006; 116 (4): 630-4.

"If you want to walk fast, walk alone; if you want to walk far, walk with friends"

Ghanain folk proverb