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Foresee Your Next Patient

PHOTOCLINIC

Laryngeal Cleft Type I

Syed A. A. Rizvi, PhD, MS, MBA¹ • Zafar Qureshi, MD² • Palvi Walia, PA-C³ • Mareena Kashif-Shafiq, BS⁴ • Rubasha Masood, BS⁵

21-month-old boy was brought to the clinic by his mother for a hospital discharge follow-up visit. The patient had a history of gastroesophageal reflux disease, asthma attack, bronchiolitis, and recurrent wheezing. He had been admitted to the hospital for respiratory distress, tachypnea, and wheezing. He had required aggressive bronchodilation with intravenous corticosteroids. The patient also had had a hospital admission 5 month prior with a similar presentation.

He had been born at 35 weeks of gestation via spontaneous vaginal delivery, and he had had a previous workup for aspiration (due to recurrent wheezing) at the age of 5 months that yielded positive bronchoalveolar lavage pepsin assay findings. His family history includes asthma in his brother, sister, mother, and paternal grandmother.

Physical examination. At presentation, the patient was awake and alert, with no distress. Movement in all 4 extremities was even, with good strength. His temperature was 37.3 °C (temporal), his blood pressure was 115/66 mm Hg, his pulse rate was 102 beats/min, and his respiratory rate was 28 breaths/min. The patient's height was 84.4 cm (50th percentile) and his weight was 10.6 kg (25th percentile). The tympanic membranes appeared normal in both ears, the oropharynx was clear, and no discharge

AFFILIATIONS:

- $^{\rm 1}{\rm Hampton}$ University School of Pharmacy, Hampton, Virginia
- ²Universal Medical Clinic, Miami, Florida
- ³Keiser University, Fort Lauderdale, Florida
- ⁴Nova Southeastern University, Fort Lauderdale, Florida
- ⁵Florida Atlantic University, Fort Lauderdale, Florida

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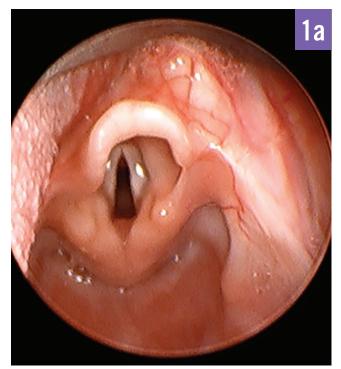
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CORRESPONDENCE:

Syed A. A. Rizvi, PhD, MS, MBA, Department of Pharmaceutical Sciences, Hampton University School of Pharmacy, 100 E Queen St, Hampton, VA 23669 (syed.rizvi@hamptonu.edu)





Endoscopic view of type 1 laryngeal cleft at presentation.





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Diagram showing the types of laryngeal cleft. Reproduced with permission from: Zucker EJ, Laya BF, Liszewski MC, Restrepo R, Vargas SO, Lee EY. Large airways. Radiology Key. Accessed December 15, 2020. https://radiologykey.com/large-airways/

was present from either eye. His hear was in regular rhythm, and his lungs were clear to auscultation with no respiratory distress. His abdomen was soft, with no distention or tenderness. His skin was warm, with no rash noted.

Clinical course. The patient had been referred to the hospital (just prior to the follow-up visit at our clinic) for direct admission after results of an outpatient swallow study showed aspiration in all consistencies. The patient had been admitted to the pediatric unit, where his case had been followed in consultation by gastroenterologists, otorhinolaryngologists, and speech therapists.

After the diagnosis of laryngeal cleft (**Figure 1**), the decision was made to operate, since there was no other reasonable explanation for his aspiration given the fact that the patient was otherwise developmentally normal. The patient met the criteria for microlaryngoscopy, bronchoscopy, and injection laryngoplasty. Surgery was performed after obtaining informed consent. Using a 24-gauge laryngeal needle, 0.4 mL of Prolaryn water-based gel was injected into the submucosa at the apex of the laryngeal cleft. There was excellent effacement of the cleft; the surgery was successful and uneventful.

Discussion. Laryngotracheal esophageal cleft or, simply, laryngeal cleft (LC), is a congenital, abnormal, posterior sagittal communication between the esophagus and the laryngotracheal complex. It is a rare condition, with a prevalence of 1 in 10,000 to 1 in 20,000 live births and with a male-to-female ratio of 5 to 3. Due to its rarity and nonspecific clinical features, it is challenging to diagnose, but with numerous options available for management ranging from conservative treatment to early surgery. 3

There are 4 types of LCs (types I through IV) based on their length as described in 1989.⁴ Type I LCs are the mildest but may not be diagnosed immediately, depending on the severity of the clinical symptoms. With types II, III, and IV, the cleft continues to extend further down the esophagus (**Figure 2**).⁵ The condition is usually associated with gastroesophageal reflux disease and genetic syndromes such as Pallister-Hall syndrome, CHARGE syndrome, VACTERL association, Opitz G/BBB syndrome, and DiGeorge syndrome.⁶

Presenting clinical symptoms include coughing, cyanosis, aspiration, and choking events, as were noted with our patient. Because neonates often show suck, swallow, and breathing incoordination,⁷ the diagnosis of LC is often missed or delayed. The diagnosis is made by direct visualization, and the gold standard examinations are microlaryngobronchoscopy and rigid laryngotracheobronchoscopy.^{3,8} Various endoscopic procedures or surgical approaches are available for the management of LC depending on the type and are often individualized.⁹

Our patient is healing well, and the plan is to follow up with an otorhinolaryngologist in 2 to 3 weeks, at which time a bedside swallow evaluation will be performed.

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