Endoscopic Surgical Repair of Type 3 Laryngeal Clefts

Eelam Adil, MD, MBA; Hasan Al Shemari, MD; Reza Rahbar, DMD, MD

Laryngeal clefts (LCs) are congenital anomalies characterized by a defect in the posterior laryngeal wall that can extend inferiorly into the trachea. The most common symptoms include aspiration, recurrent pneumonia, cyanotic episodes, and difficulty feeding. Diagnosis requires a high index of suspicion based on clinical presentation, interpretation of preoperative studies, and a thorough endoscopic evaluation using general anesthesia.1,2

The most commonly used classification system was described by Benjamin and Inglis3 in 1989. In this system, type 1 clefts are interarytenoid defects that extend to the level of the true vocal folds. Type 2 clefts extend below the level of the true vocal folds, through a portion of the posterior cricoid lamina. Type 3 clefts extend completely through the cricoid cartilage, with or without extension into the cervical tracheoesophageal wall, and type 4 clefts extend through the entire length of the tracheoesophageal wall into the thorax. Type 3 laryngeal clefts (LC type 3) are very rare and their repair is commonly approached through an open technique.4 The aim of this study was to describe our surgical technique with a focus on when we consider endoscopic repair feasible. The clinical outcome of endoscopic carbon dioxide–assisted cleft repair in these patients is also reviewed.

Methods

After obtaining approval from the institutional review board of Boston Children’s Hospital, a retrospective medical record review study was performed on all patients with LC type 3 diagnosed via direct laryngoscopy and rigid bronchoscopy from January 2007 to September 2013 at a tertiary pediatric hospital.
A total of 6 patients were identified. No patient was excluded from the study, and all underwent endoscopic repair. Two patients had been transferred from outside institutions after the diagnosis of LC type 3 was confirmed.

To determine the type of cleft, a direct laryngoscopy is performed using a Parson laryngoscope and 4-mm, 0° telescope. The interarytenoid area is exposed using a laryngeal spreader. The interarytenoid area is then palpated using a probe to confirm the diagnosis and measure the length of the cleft (Figure, A, J, N, and P). Laryngeal cleft type 3 is diagnosed when the cleft extends completely through the cricoid cartilage. The whole procedure is performed with the patient spontaneously ventilating.

All of these patients underwent endoscopic carbon dioxide laser-assisted repair of their LC. Endoscopic repair was performed using suspension microlaryngoscopy with the administration of general anesthesia with spontaneous ventilation using the anesthesia technique we previously described. The larynx was visualized with a Lindholm laryngoscope. An OmniGuide Surgical carbon dioxide laser at a setting of 6 W at 0.3- to 0.4-second intermittent mode was used to denude the mucosal lining beginning at the apex of the cleft and extending cranially (Figure, B, H, K, and Q). Curved fine microlaryngeal...
alligator forceps were used to spread the mucosa during ablation with the laser. Cotton pledgets soaked with oxytetracycline hydrochloride (Afrin; Merck) were applied to the area for hemostasis. It is important to remove the mucosa completely at the apex of the cleft to prevent development of a fistula at the distal end of the repair. Absorbable interrupted sutures (4-0, 5-0, and/or 6-0 Vicryl on P1 or P3 needles; Ethicon Inc) were used to reapproximate the mucosal edges (Figure, R). The first suture was the most important and placed at the most caudal extent of the cleft. We generally placed 3 to 4 sutures in a distal to proximal fashion, depending on the extent of the cleft. The first suture was approximately at the apex of the cleft, the second was placed slightly below the first suture, and the remaining sutures were placed progressively inferior to the previous suture.

Following cleft repair, the patients were transferred to the pediatric intensive care unit (PICU) for observation. Patients are typically monitored in the PICU overnight and then transferred to floor status for observation for another 48 hours. They resume their preoperative diet and receive dexamethasone (0.5 mg/kg, up to 10 mg) for 24 hours. Following discharge, they are seen in clinic 1 week later for flexible fiberoptic laryngoscopy to examine their surgical site. A postoperative modified barium swallow (MBS) is performed 3 months following surgery to reevaluate their swallow function.

Outcome measures including demographics, medical comorbidity, swallowing outcome, and complications were collected and analyzed. Descriptive statistics were calculated to describe the overall group.

**Results**

Over the study period, 6 patients were diagnosed as having LC type 3 and treated with endoscopic carbon dioxide laser-assisted repair (Table 1). The median age at diagnosis was 4 months (interquartile range [IQR], 1.6 months) and at endoscopic surgical repair, 7.5 months (IQR, 2.1 month). There was a male to female ratio of 5:1. Congenital anomalies were found in 4 patients (67%); 2 had VACTERL association (vertebral, anal, cardiac, tracheal, esophageal, renal, and limb anomalies), 1 had isolated tracheoesophageal fistula (H-type), and 1 had Opitz syndrome. In 4 patients (67%), the cleft extended through the cricoid cartilage and ended above the first tracheal ring. In 1 patient, the cleft passed to the first tracheal ring and in the last patient the cleft extended to the second tracheal ring.

At presentation, recurrent aspiration pneumonia was the chief complaint in 4 patients (67%). Two patients (33%) presented with feeding difficulty and weight loss. All patients had aspiration of thickened liquids documented by MBS before the endoscopic repair indicating severe aspiration and precluding an oral diet. Median operative time was 98.2 minutes (IQR, 16.0 minutes). Five patients (83%) underwent gastrostomy tube placement, and 1 had a nasogastric tube for feeding prior to laryngeal cleft repair. Nissen fundoplication was performed on 2 patients (33%).

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**Table 1. Characteristics of 6 Patients With Type 3 Laryngeal Clefts and Treated With Endoscopic Carbon Dioxide Laser–Assisted Repair**

<table>
<thead>
<tr>
<th>Characteristic</th>
<th>Patient No.</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sex</td>
<td>Male</td>
</tr>
<tr>
<td>Age at first presentation, mo</td>
<td>1.9</td>
</tr>
<tr>
<td>Chief complaint</td>
<td>Feeding difficulty with weight loss</td>
</tr>
<tr>
<td>Age at endoscopic repair, mo</td>
<td>6.9</td>
</tr>
<tr>
<td>Syndrome or congenital anomalies</td>
<td>None</td>
</tr>
<tr>
<td>Aspiration-related procedures</td>
<td>Gastrostomy, fundoplication</td>
</tr>
<tr>
<td>Comorbidities</td>
<td>None</td>
</tr>
<tr>
<td>Tracheostomy</td>
<td>No</td>
</tr>
<tr>
<td>MBS Before repair</td>
<td>Thickened fluid aspiration</td>
</tr>
<tr>
<td>After repair</td>
<td>No aspiration</td>
</tr>
<tr>
<td>Extent of the cleft</td>
<td>Above first tracheal ring</td>
</tr>
<tr>
<td>Second revision endoscopic repair</td>
<td>No</td>
</tr>
</tbody>
</table>

Follow-up, mo 12 13 48 16 12 5

Abbreviations: ASD, atrial septal defect; EA, esophageal atresia; IVH, intraventricular hemorrhage; MBS, modified barium swallow; NA, not available; TEF, tracheoesophageal fistula; VACTERL, vertebral, anal, cardiac, tracheal, esophageal, renal, and limb anomalies; VC, vocal cord.

* Presented to our institution at 32 months.
had mixed results (described in the literature, but endoscopic repair of LC type 3 has successful endoscopic repair of LC types 1 and 2 has been de-

revision procedures. Thiel et al\textsuperscript{10} also presented 1 case of LC acquired a second endoscopic procedure to correct small, re-

tempted in 4 patients with type 3 clefts. Two patients re-

quired a second endoscopic repair in 4 patients with type 3 clefts (Table 2).\textsuperscript{3,4,11} Sandu et al\textsuperscript{7} showed the suc-

cessful use of endoscopic repair in 4 patients with type 3 clefts without any complications. All 4 patients in their series achieved normal feeding with no clinical signs of aspiration and all had a good voice following surgery. However, they ex-
cluded patients with secondary airway abnormalities (laryngomalacia, tracheomalacia, tracheoesophageal fistula) and as-

iated congenital anomalies (Pallister-Hall and Opitz syndromes).\textsuperscript{7}

Garabedian et al\textsuperscript{8} also reported results of endoscopic re-

pair in 4 patients with LC type 3. They included 2 patients with congenital anomalies (pyelocalyceal dilatation and Opitz syn-

drome), and both required a second endoscopic procedure. In the series by Broomfield et al,\textsuperscript{9} endoscopic repair was at-

tempted in 4 patients with type 3 clefts. Two patients re-

quired a second endoscopic procedure to correct small, re-

tidual type 1 defects, and the other 2 patients required open revision procedures. Thiel et al\textsuperscript{10} also presented 1 case of LC type 3b in their series in whom both endoscopic and subse-

quent open repair approaches had failed.

The endoscopic approach for the repair of LCs has several potential advantages over open approaches. Endoscopic re-

pair avoids some of the risks of the external approach including wound complications and laryngeal airway instability fol-

lowing laryngofissure. In addition, if a lateral pharyngotomy approach is selected for open repair, there is potential for in-

jury to the recurrent laryngeal nerve. Furthermore, endo-

scopic repair may avoid tracheal intubation and tracheostomy.\textsuperscript{7} Finally, patients who undergo endoscopic repair potentially have a shorter hospital stay. In our series, 5 patients (83%) were discharged after 3 days.

In our experience, there are 2 factors that determine when endoscopic repair is possible. First, the ability to maintain an-

esthesia with spontaneous ventilation is necessary for endo-

scopic repair. Prior to considering an endoscopic approach, a conversation with the anesthesiologist is necessary to deter-

mine if this will be possible from a cardiopulmonary perspec-

tive. The median operative time in our series was 98.2 min-

utes (IQR, 16 minutes) which means the anesthesia team must be comfortable with maintaining the patient at this anesthe-

sia level for 1.5 to 2.0 hours.

If the anesthesiologist is confident that the appropriate an-

esthesia level can be maintained for the duration of the pro-

cedure, then the second requisite to an endoscopic approach is adequate posterior glottic exposure. The depth of the cleft has to be visible and accessible for suture placement while the patient is in suspension. At the depth of the cleft where ac-

cess is most limited, we find it useful to use 5-0 or 6-0 Vicryl sutures on a P1 needle. As repair proceeds cranially, we in-

crease the size of the suture and needle to 4-0 or 5-0 Vicryl on a P3 needle.

In our series, the median age at diagnosis was 4 months (IQR, 1.6 months) and at endoscopic surgical repair, 7.5 months (IQR, 2.1 month). The apparent lapse between diagnosis and repair is largely to optimize their pulmonary status prior to sur-

ery. As mentioned previously, all of these patients aspirate and have varying degrees of lung injury and/or infection as a result. These issues are challenging and can delay both endo-

scopic and open repair. Therefore, we often refer these pa-

tients to our pulmonary colleagues to optimize their pulmo-

nary function prior to surgery. Bronchodilators, respiratory synctial virus prophylaxis, and antibiotics are often neces-

sary regardless of approach.

One of the goals of endoscopic repair is to avoid intuba-

tion and tracheotomy placement. When repairing a LC type 3, it is important to remember that as the cleft is closed, the air-

way is being narrowed. In addition, manipulation of the air-

way will cause some swelling. Therefore, we tend to be cau-

tious with our endoscopic repairs and would prefer to place less sutures and possibly return to the operating room for end-

oscopic repair of a residual defect rather than risk post-

operative airway compromise that may require intubation and/or tracheotomy placement. There was 1 patient in our se-

Table 2. Summary of Prior Reports of Type 3 Laryngeal Clefts

<table>
<thead>
<tr>
<th>Source</th>
<th>Patients, No.</th>
<th>Primary Surgery Success Rate, %</th>
<th>No. Who Underwent Second Endoscopic Procedure</th>
<th>Secondary Endoscopic Success Rate, %</th>
<th>Comorbidities in Primary Repair Failures</th>
</tr>
</thead>
<tbody>
<tr>
<td>Sandu et al\textsuperscript{7}</td>
<td>4</td>
<td>100</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>Garabedian et al\textsuperscript{8}</td>
<td>4</td>
<td>50</td>
<td>2</td>
<td>100</td>
<td>Pyelocalyceal dilatation, Opitz syndrome</td>
</tr>
<tr>
<td>Broomfield et al\textsuperscript{9}</td>
<td>4</td>
<td>0</td>
<td>2</td>
<td>100</td>
<td>NA</td>
</tr>
<tr>
<td>Thiel et al\textsuperscript{10}</td>
<td>1</td>
<td>0</td>
<td>0</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>

Abbreviations: LC, laryngeal cleft; NA, not available.
ries who required a second endoscopic procedure for repair of a symptomatic residual type 1 cleft. Aspiration completely resolved following the second procedure.

Patient number 3 in our series required a tracheotomy. This was performed 2.5 months before the diagnosis of LC type 3. The tracheotomy was not related to LC type 3 or to the endoscopic cleft repair. It was due to prolonged intubation and failed extubation after tracheoesophageal fistula repair. She was decannulated 4 months ago.

Four patients in our series had congenital anomalies or syndromes. We had 2 patients with VACTERL who also had vocal cord paralysis following tracheoesophageal fistula repair. Patient 3 had no aspiration on her first postoperative MBS. She currently takes half of her feedings by mouth and half via G-tube because she has severe oral aversion and microgastria that continue to limit her oral intake. Patient 6 was our most recent repair, and his 3-month follow-up MBS showed aspiration with nectar thickened liquids, but no aspiration with honey-thickened liquids or purees. He has oropharyngeal dysphagia characterized by reduced oral motor skills and delayed onset of the swallow as well as persistent left vocal fold immobility, which are likely the cause of his aspiration. He currently takes purees by mouth and supplements with G-tube feeds. He has not had any episodes of aspiration pneumonia since his cleft repair. This indicates that endoscopic carbon dioxide laser-assisted repair of LC type 3 is a valuable procedure even in patients with significant comorbidities.

Conclusions

The management of LC remains challenging, despite developments in surgical instrumentation and technique. We have noticed a trend toward fewer failed repairs possibly because of our extensive experience with endoscopic repair of LC types 1 and 2. The ability to maintain an adequate level of anesthesia and sufficient posterior glottic exposure are the 2 factors we consider when determining if a patient is a candidate for endoscopic repair. Overall, the outcomes in our series of LC type 3 have been favorable with no major complications. We believe that the endoscopic carbon dioxide laser-assisted procedure is a relatively safe and reproducible technique for repair of LC type 3.

REFERENCES