Does the Presence of a Tracheoesophageal Fistula Predict the Outcome of Laryngeal Cleft Repair?

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Objective: To determine if the presence of a tracheoesophageal fistula (TEF) alters outcome following laryngeal cleft repair.

Design: A retrospective review of patients diagnosed and treated for laryngeal clefts, with a minimum follow-up period of 1 year.

Setting: An academic tertiary care children's hospital.

Patients: Twenty-five pediatric patients diagnosed and surgically treated for laryngeal cleft.

Main Outcome Measures: Each chart was reviewed to determine if patients with a laryngeal cleft had been diagnosed with TEF and had undergone a surgical TEF repair procedure. The success of the surgery was evaluated based on the resolution of symptoms and the endoscopic evaluation of the repair site.

Results: Twenty-five patients were reviewed for study purposes. Fourteen had a history of TEF repair and 11, no history of TEF. All 25 patients underwent surgical repair of the laryngeal cleft. Twelve of the 14 patients with a history of TEF repair experienced a breakdown of the laryngeal cleft repair. Only 1 of the 11 patients with no history of TEF experienced such a breakdown. In 8 of 9 patients with a laryngotraheoesophageal type I cleft, surgical repair was not successful.

Conclusions: In our series, patients with laryngeal clefts who also had a history of TEF had a much higher incidence of failed cleft repair compared with patients with no history of TEF. This finding is not conclusive and requires further investigation. The failure of cleft repair correlated with the severity of the cleft. The importance of these associations may lead to enhanced surgical planning and realistic preoperative family expectations.


Laryngeal clefts (LCs) and laryngotraheoesophageal clefts (LTECs) are rare congenital anomalies. The approximate incidence is 1 in 10,000 to 20,000 live births.1

Multiple congenital anomalies can be associated with LC and LTEC. These include tracheoesophageal fistulas (TEFs), laryngomalacia, cleft lip and palate, subglottic stenosis, hypoplastic lungs, transposition of the great vessels, hamartoma, imperforate anus, rectal stenosis, mesenteric malrotation, Meckel diverticulum, hypoplasia, and hypoplastic kidneys.2

Tracheoesophageal fistulas have been identified in 20% to 37% of patients with LC and LTEC.3,4 Two senior authors (R.T.C. and C.M.M.) have observed that a much higher incidence of failed LC and LTEC repairs can be found in patients with a history of TEF. The purpose of this study was to specifically compare the surgical outcome of LC and LTEC repair in patients with and without a history of TEF.

A major difficulty in discussing the surgical options and results in patients with airway clefts is the multiple classification schemes and confusing nomenclature used. Classification schemes have been described by Pettersson, Armitage, Benjamin, Evans, Myer, and others. To discuss surgical technique or surgical outcome, one must compare clefts involving the same anatomical regions. For this reason, we use the Myer-Cotton5 scheme, outlined below, which is anatomically specific and descriptive.

<table>
<thead>
<tr>
<th>Cleft Location</th>
<th>Cleft Type</th>
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<tbody>
<tr>
<td>Laryngeal</td>
<td></td>
</tr>
<tr>
<td>Interarytenoid</td>
<td>L I</td>
</tr>
<tr>
<td>Partial cricoid</td>
<td>L II</td>
</tr>
<tr>
<td>Complete cricoid</td>
<td>L III</td>
</tr>
<tr>
<td>Laryngotraheoesophageal</td>
<td>LTE I</td>
</tr>
<tr>
<td>Into trachea</td>
<td></td>
</tr>
<tr>
<td>To carina</td>
<td>LTE II</td>
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</table>

This system differs from most others by dividing the clefts into LCs and LTECs rather than attempting to use one term to describe all variations. Each of these is then subdivided to give specific information as to the location of the cleft.
PATIENTS AND METHODS

We performed a retrospective review of patients diagnosed and treated for LC and LTEC between January 1985 and December 1996 at Children's Hospital Medical Center, Cincinnati, Ohio. Inclusion in our review required a minimum of 1-year follow-up from the time of surgical repair. Patients with L I– or LTE II–type clefts were excluded from the study. All charts were reviewed and data collected regarding age at the time of cleft repair, the presenting symptoms, the type of cleft present (based on the Myer-Cotton classification system), any associated anomalies or comorbid conditions, the type of cleft repair performed, and the postoperative status with regard to the formation of a recurrent cleft or an intact surgical repair site.

RESULTS

Twenty-five patients' charts were available for review. The average age at the time of surgery was 2.4 years (age range, 2 weeks to 9 years). The most common presenting symptoms were feeding problems, aspiration, stridor, and cyanotic episodes. The associated anomalies and comorbidities are listed below.

Anomalies and/or Comorbidities Patients, No. (%)  
- Tracheomalacia 15 (60)  
- Esophageal atresia 7 (28)  
- Subglottic stenosis 6 (24)  
- Bronchomalacia 5 (20)  
- Congenital heart disease 4 (16)  
- Opitz syndrome 4 (16)  
- Prematurity 4 (16)  
- Hypospadias 4 (16)  
- Anal stenosis 3 (12)  
- Polyhydramnios 3 (12)  
- Vater syndrome 2 (8)  
- Malariaion of gut 2 (8)  
- Bronchopulmonary dysplasia 2 (8)

Fifteen of the patients required preoperative feeding tubes, and 11 underwent preoperative Nissen fundoplications. Twenty patients had tracheostomy tubes placed preoperatively, and 14 were decannulated at some point in the first year following surgery. Six patients required a laryngotracheoplasty with costal cartilage grafting prior to eventual decannulation. Three patients had redundant mucosa in the posterior aspect of the airway following repair; 2 of these required laser excision of the mucosa. Two patients died approximately 1 year following surgery, only 1 whom had an intact surgical repair site.

Fourteen patients had a history of TEF and had undergone TEF repair prior to the LC or LTEC surgery. Twelve (86%) of these patients developed a recurrent LC or LTEC. Seven of these patients required multiple (>3) procedures in the attempt to repair the cleft.

Eleven patients had no history of TEF. One (9%) of these patients developed a recurrent LC or LTEC.

Cleft types are listed in the Table. All cleft procedures (LC and LTEC) were performed using an anterior laryngotomy. Two patients had undergone initial repair at other institutions for which a lateral pharyngotomy was used. In 8 of 9 patients with LTE I clefts, breakdown of the surgical repair was identified.

<table>
<thead>
<tr>
<th>Cleft Type</th>
<th>TEF Group</th>
<th>Non-TEF Group</th>
</tr>
</thead>
<tbody>
<tr>
<td>L I</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>L II</td>
<td>3 (2/3 recurred)</td>
<td>5 (0/5 recurred)</td>
</tr>
<tr>
<td>L III</td>
<td>3 (3/3 recurred)</td>
<td>5 (0/5 recurred)</td>
</tr>
<tr>
<td>LTE I</td>
<td>8 (7/8 recurred)</td>
<td>1 (1/1 recurred)</td>
</tr>
<tr>
<td>LTE II</td>
<td>0</td>
<td>0</td>
</tr>
</tbody>
</table>

* TEF indicates tracheoesophageal fistula; L, laryngeal; and LTE, laryngotracheoesophageal.

Laryngeal clefts and LTECs are uncommon congenital anomalies. Many associated conditions can coexist, including congenital heart disease, gastrointestinal and gastrourinary tract abnormalities, and midline defects. The condition is thought to be familial, with an autosomal dominant inheritance pattern. In addition, LC and LTEC are identified as part of 2 syndromes: the Opitz-Frias syndrome, or G syndrome, consists of hypertelorism, cleft lip, cleft palate, airway cleft, and hypospadias; the Pallister-Hall syndrome consists of congenital hypothalamic hamartoblastoma, hypopituitarism, airway cleft, imperforate anus, and postaxial polydactyly.

Pettersson performed the first surgical repair of an LC in 1955. Postsurgical success of LCs depends on maintaining an intact wall separating the airway and the digestive tract. Evans et al described 15 patients who had open repairs of LCs or LTECs, 4 (26.7%) of which required revision. Two of the repairs required multiple revisions, and 1 patient died prior to revision surgery. Other studies have reported anastomotic leaks in as many as 50% of postoperative patients. A review of 170 clefts reported prior to 1991 identified only 19 (11.2%) that required revision. Most studies, including ours, have concluded that the incidence of revision surgery increases with the severity of the cleft.

Treatment options depend on the classification of the cleft. Type L I clefts may be treated in some cases with antireflux therapy alone. If still symptomatic, these patients may need an endoscopic or open surgical repair. Types L II, L III, and LTE I are best treated using an anterior laryngotomy. Type LTE II requires a lateral pharyngotomy. Most patients will require a tracheostomy either preoperatively or peroperatively. For shorter-length clefts that require only minimal airway manipulation, a single-stage approach may be reasonable. For more extensive clefts, a tracheostomy will likely be necessary for airway control and protection.

Our study found a much higher incidence of breakdown of cleft repair in patients with a history of TEF. Though most of these patients had undergone their initial repair at other institutions, the referral pattern appeared to be spread equally throughout the country. The
The common type of TEF is esophageal atresia with a distal TEF, the esophageal gap usually being 2 cm in length and the fistula most often occurring at the carina area. This type of defect is found in 85% of patients with TEF. The H type of TEF accounts for only 3% to 5% of all TEFs. In our 14 patients with TEF, only 7 had esophageal atresia. In other studies on patients with TEF, the incidence of LC or LTEC is only about 6%. In patients with both LTEC and TEF, cine-esophagograms may show rapid "spillover" into the trachea with less than 1 mL of contrast material. Endoscopy remains the mainstay in diagnosing LTEC. This should consist of a micro-laryngoscopy, bronchoscopy, and esophagoscopy. Often an anterior commissure–type laryngoscope is helpful, along with a suspension apparatus to provide the best visualization of the posterior laryngeal and tracheal region. The condition involves inolding of tissue in the posterior aspect of the airway, which must be splayed to its maximum width to fully evaluate the cleft. It is essential to identify the length of the cleft and its relationship to key structures, including the true vocal folds, the cricoid cartilage, and the carina. A narrow flexible ureteric catheter can be used to palpate the posterior tracheal wall and probe previously repaired TEF sites to ensure that they are intact. It is not uncommon to find a small pouch at the TEF repair site, which may have a more funicular shape in a patient with LTEC than in one without a cleft.

A stable airway must be established for each patient and aspiration prevented. Chronic and irreversible pulmonary injury can occur from a few weeks of aspiration, especially in the neonate. Gastroesophageal reflux must be controlled prior to surgery to optimize the cleft repair.

We believe that the anterior laryngotomy is best for all type I, II, I, III, and LTE I clefts. This approach offers excellent exposure of the cleft. Because it is performed in the midline, it avoids injury to vascular and neural structures. In our experience, careful reapprroximation of the airway does not cause permanent sequelae. We use a 2-layer closure with absorbable sutures. If possible, the 2 layers should not directly overlay each other. If the tissue appears weak in a primary or secondary cleft repair, a sternocleidomastoid or strap muscle flap (inferiorly based) must be positioned between the 2 tissue layers for added strength. Recurrent cleft repair generally necessitates complete fasciectomy of the tissue from the cleft site superiorly to the interarytenoid region to allow for a smooth suture line. If subglottic stenosis is present, a laryngotracheoplasty with or without costal cartilage grafting may be needed in addition to the cleft repair. Caution must be exercised if using a posterior graft in patients with a deficient posterior cricoid ring. We find it helpful to place a Maloney-type esophageal dilator in the esophagus (transoral) prior to the procedure to assist with differentiating esophageal mucosa and estimating the amount of esophageal mucosa to preserve once the cleft site is exposed transcervically.

Our study excluded patients with type L I clefts because many of these do not require surgery. When surgery is required, it is controversial whether the approach should be endoscopic or open. We also excluded type LTE II clefts because these are often acutely life threatening and involve an extensive care and decision making process. Discussion of these 2 excluded groups is beyond the scope of this article.

Recurrent LTECs are thought to occur more commonly when the primary repair used nonabsorbable suture material and when the suture lines are in direct apposition. The use of muscle interposition with the sternohyoid muscle was described in 1976 to treat 2 recurrent clefts. Techniques have also been described using other material for interposition. We use muscle interposition to enhance the integrity of the surgical repair for primary cases of poor-quality tissue and for recurrent cleft repairs.

Our intention is to make pediatric otolaryngologists and surgeons aware of the poor outcome of LC and LTEC repair in patients with a history of TEF. This information may lead us to using muscle interposition at the time of primary cleft repair in patients with TEF. Close follow-up is essential in the TEF group of patients following cleft repair. Preoperative parental meetings need to address the issue of follow-up and potential breakdown of cleft repair.

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REFERENCES