Laryngomalacia: A classification system and surgical treatment strategy

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Abstract
Laryngomalacia, the most common congenital laryngeal anomaly, is not a single disease entity but rather a variety of entities along a spectrum of underlying pathophysiological processes. Based on our study of 10 children who were surgically treated for laryngomalacia in an urban tertiary care center, we have developed a system of classifying laryngomalacia on the basis of its different underlying pathophysiologic processes. Type 1 laryngomalacia is characterized by a foreshortened or tight aryepiglottic fold. Type 2 disease is defined by the presence of redundant soft tissue in the supraglottis. The type 3 designation applies to cases caused by other etiologies, such as underlying neuromuscular disorders. While the three types are not mutually exclusive, each should be considered as a separate disease entity with a final common clinical presentation. Each type requires a specific approach to surgical repair.

Introduction
Laryngomalacia is the most common cause of neonatal stridor, accounting for one-half to two-thirds of all cases of noisy breathing in this age group. Laryngomalacia usually becomes symptomatic during the first 2 weeks of life, and it usually resolves by the time a child is 12 to 18 months old. Only 10% of cases require surgical intervention, generally to treat apnea or failure to thrive. The underlying defect that causes laryngomalacia remains unknown, but it is believed to involve neuromuscular weakness, cartilaginous inadequacy, or anatomic abnormality such as redundant arytenoid mucosa.

Various classification schemes for laryngomalacia have been proposed, and assorted treatment modalities and surgical procedures have been described. However, few authors have attempted to mesh the classifications and their treatments to develop a treatment algorithm. In this article, we describe our efforts to tailor the surgical procedure according to the specific type of laryngomalacia.

Patients and methods
We established a hypothesis that the surgical treatment of laryngomalacia specifically directed at the underlying etiology would be effective and sufficient. To test our theory, we developed a classification system for laryngomalacia based on its underlying etiology (figure), and then we determined the most appropriate surgical procedure for each diagnosis.

Type 1 laryngomalacia is characterized by a foreshortened or tight aryepiglottic fold, and we treated it by dividing the folds with scissors. Type 2 disease is defined by the presence of redundant soft tissue in the supraglottis, and it was treated by resecting the redundant mucosa with sharp instruments. The type 3 designation applied to cases caused by other etiologies, such as underlying neuromuscular disorders and posterior epiglottic collapse; it was treated with a tracheostomy.

We studied our hypothesis in 10 patients—8 boys and 2 girls aged 3 months to 3 years (median: 6 mo).

Results
Two of the 10 patients had been born prematurely with bronchopulmonary dysplasia and gastroesophageal reflux (table). The most common reason for surgical intervention was respiratory distress (n = 8), defined as worsening stridor and retraction beyond what is normally seen in laryngomalacia. Six patients experienced failure to thrive, 3 had apnea, and 2 had cyanosis.

Five patients had isolated type 1 laryngomalacia, 3 had isolated type 2 laryngomalacia, 1 patient had a combination of types 1 and 2, and 1 patient had a combination of types 1 and 3. No patient had isolated type 3 laryngomalacia. Other findings noted at the time of direct laryngoscopy and bronchoscopy included posterior laryngeal edema in 4 patients and subglottic stenosis in 2 patients.
Supraglottoplasty was performed on 9 patients; 6 patients underwent an aryepiglottic fold division, and 4 underwent an arytenoid mucosal resection (1 patient underwent both procedures). Tracheostomy was performed on 2 patients, once as a primary procedure and once following the failed resection of bilateral arytenoid redundant mucosa.

Discussion
The three types of laryngomalacia we describe do not represent a novel classification system. In fact, our classifications reflect the three abnormalities that McSwiney et al in 1977 noted could be present, either separately or in combination. They wrote that (1) the epiglottis could be long and curled upon itself and prolapse posteriorly on inspiration, (2) the aryepiglottic folds could be too short, and (3) the arytenoids could be more bulky than normal and prolapse forward on inspiration. They also noted a high incidence of laryngomalacia among children with cerebral palsy and mental retardation, although they did not specify the particular type of laryngomalacia in these particular patients. In our experience, patients with neurodevelopmental disorders generally have a posterior prolapsed epiglottis; when surgical intervention is required in such cases, tracheostomy rather than supraglottoplasty is the procedure of choice.

Other authors have advocated different approaches to the surgical treatment of laryngomalacia. In a series of 40 surgical cases of laryngomalacia, Prescott advised surgeons to focus on three supraglottic elements during the physical examination: the epiglottis, the aryepiglottic folds, and the mucosa over the corniculate cartilages. He found an omega-shaped epiglottis in 27 patients (67.5%), with epiglottal prolapse into the laryngeal inlet in 13 patients (32.5%). All patients had a short aryepiglottic fold and edematous mucosa over the corniculate cartilages that prolapsed anteriorly. Prescott advocated excision of a V wedge from the center of the aryepiglottic folds as the surgical procedure of choice. He reported resecting the mucosa over the corniculate cartilages only if it was edematous, and in fact he did perform this procedure on every one of the patients he described in that study. He excised the lateral margins of an omega-shaped epiglottis in 32 patients (80.0%) and noted that no patient had persistent epiglottic prolapse requiring epiglottoplasty.

Nussbaum and Maggi defined two types of laryngomalacia based on criteria important to pulmonologists. The first type is isolated laryngomalacia as defined by McSwiney et al, who noted that this entity is usually detected in early infancy. The second type is laryngomalacia associated with any other bronchoscopic findings, including gastroesophageal reflux.

In describing a series of 115 patients, Roger et al distinguished three types of laryngomalacia: complete, predominantly posterior, and isolated anterior. The complete type, which was seen in two-thirds of their patients, was characterized by an omega-shaped epiglottis, shortened aryepiglottic folds, and redundant supraglottic mucosa that may or may not involve the mucosa of the cuneiform cartilages. Most of the other patients had the predominantly posterior type with redundant arytenoid mucosa. Only 2 patients had isolated anterior laryngomalacia with posterior swaying of the epiglottis.

Shah and Wetmore, in designing a reporting form for laryngomalacia, separated the disorder into three types based on the principal site of anatomic collapse. In their series of 10 patients, 4 had posterolateral laryngomalacia with redundant aryepiglottic folds, and 6 had only posterior laryngomalacia with excess arytenoid mucosal or cartilaginous bulk; no patient had anterior laryngomalacia.
involving the epiglottis. Two of their patients required supraglottoplasty; the laryngomalacia type and surgical technique in these 2 cases were not specified.

Another classification system—in which laryngomalacia is designated as type A, B, or C—is cited in the Taiwanese literature. In type A disease, the cuneiform cartilages are redundant and they prolapse during inspiration. In type B laryngomalacia, a long tubular epiglottis curls back upon itself during inspiration. In type C, the flaccid epiglottis prolapses posteriorly against the posterior pharyngeal wall or vocal folds during inspiration.

Nielson et al developed an empiric scale of 0 to 8 for rating laryngomalacia. The arytenoids and the epiglottis are graded separately on a scale of 0 to 4 points, and the grades are added together. The arytenoids may show no collapse (0), subtle collapse (1), collapse that obscures 25 to 50% of the true vocal folds (2), collapse that obscures 75% of the true vocal folds (3), or collapse that obscures 100% of the true vocal folds (4). The epiglottis may show no folding (0), slight lengthwise folding (1), moderate folding without contact of the lateral edges of the epiglottis (2), folding with intermittent contact of the lateral edges (3), or folding with continuous contact with or overlap of the lateral edges (4). This scale provides a potential

<table>
<thead>
<tr>
<th>Age/sex</th>
<th>Birth history</th>
<th>Medical history</th>
<th>Surgical indication</th>
<th>LM type</th>
<th>Other findings</th>
<th>Procedure</th>
</tr>
</thead>
<tbody>
<tr>
<td>3 mo/M</td>
<td>Normal</td>
<td>FTT</td>
<td>1</td>
<td>Resection of bilateral AE folds</td>
<td></td>
<td></td>
</tr>
<tr>
<td>8 mo/M</td>
<td>Normal</td>
<td>GER</td>
<td>FTT, respiratory distress</td>
<td>1</td>
<td>Resection of bilateral AE folds</td>
<td></td>
</tr>
<tr>
<td>3 yr/M</td>
<td>Normal</td>
<td>Hypotonia</td>
<td>Respiratory distress</td>
<td>1</td>
<td>Resection of bilateral AE folds</td>
<td></td>
</tr>
<tr>
<td>5 mo/M</td>
<td>Normal</td>
<td>FTT</td>
<td>1</td>
<td>Resection of bilateral AE folds</td>
<td></td>
<td></td>
</tr>
<tr>
<td>4 mo/M</td>
<td>Normal</td>
<td>Respiratory distress</td>
<td>1</td>
<td>SS</td>
<td>Resection of bilateral AE folds</td>
<td></td>
</tr>
<tr>
<td>7 mo/M</td>
<td>Normal</td>
<td>FTT, respiratory distress, apnea</td>
<td>1, 2</td>
<td>PLE</td>
<td>Resection of bilateral AE folds, resection of unilateral redundant mucosa</td>
<td></td>
</tr>
<tr>
<td>7 mo/M</td>
<td>Normal</td>
<td>Respiratory distress</td>
<td>2</td>
<td>SS</td>
<td>Resection of unilateral redundant mucosa</td>
<td></td>
</tr>
<tr>
<td>4 mo/F</td>
<td>Normal</td>
<td>Respiratory distress, apnea</td>
<td>2</td>
<td>PLE</td>
<td>Resection of unilateral redundant mucosa</td>
<td></td>
</tr>
<tr>
<td>4 mo/M</td>
<td>Premature</td>
<td>BPD, GER</td>
<td>FTT, respiratory distress, cyanosis</td>
<td>2</td>
<td>PLE</td>
<td>Initial: resection of bilateral redundant mucosa; later: tracheostomy</td>
</tr>
<tr>
<td>3 yr/F</td>
<td>Premature</td>
<td>MRCP, BPD, GER</td>
<td>FTT, respiratory distress, cyanosis, apnea</td>
<td>1, 3</td>
<td>PLE</td>
<td>Tracheostomy</td>
</tr>
</tbody>
</table>

LM = laryngomalacia; FTT = failure to thrive; AE = aryepiglottic; GER = gastroesophageal reflux; SS = subglottic stenosis; PLE = posterior glottic edema; BPD = bronchopulmonary dysplasia; MRCP = mental retardation and cerebral palsy.
measure of clinical severity that can complement other systems that classify specific anatomic locations such as the aryepiglottic folds.

In a comprehensive review of laryngomalacia published in 1999, Olney et al classified both surgical and nonsurgical cases into three types. Type 1 laryngomalacia was characterized by prolapse of the mucosa overlying the arytenoid cartilages, type 2 involved foreshortened aryepiglottic folds, and type 3 involved posterior displacement of the epiglottis. Types 1, 2, and 3 laryngomalacia were identified in 57, 15, and 13% of their patients, respectively; another 15% had a combination of types, usually types 1 and 2. Our system is similar to theirs, which they established midway through their study about the same time that we created our system. Just as we did, Olney et al used their classification system to direct their type of supraglottoplasty they performed. Type 1 patients underwent excision of the redundant mucosa over the posterolateral arytenoids, type 2 patients underwent division of the aryepiglottic folds, and type 3 patients underwent epiglottopexy to the base of the tongue. The surgical success rate was 78%. In our series and in the series by Olney et al, all surgical patients who had posterior epiglottic prolapse (type 3) ultimately required a tracheostomy to secure the airway. Also in our series, both of the patients with bronchopulmonary dysplasia required a tracheostomy.

The development of our classification system was motivated by our belief that different anatomic etiologies of laryngomalacia should be treated with different surgical procedures. In many previous reports, authors advocated that all surgical laryngomalacia be treated the same way, regardless of etiology. We believe that such a strategy carries the potential for unnecessary interventions.

Some authors advocate dividing only the aryepiglottic folds, be it with sharp instrumentation or with a laser. In a series of 115 patients, Garabedian et al reported a 98% success rate, with 7 patients requiring additional procedures and 2 failures requiring tracheostomy. On the other hand, some authors advocate resecting only the redundant mucosa. For example, Polonovski et al designed a suction test that involved placing an aspiration cannula into the laryngeal inlet to assess the amount of supraglottic collapse and to ascertain how much and which tissue to resect. While they frequently resected mucosa from the aryepiglottic fold and occasionally from the lateral edge of the epiglottis, they did not divide the aryepiglottic fold, claiming that two-thirds of patients who undergo such dissection eventually require a revision procedure. Still other authors advocate both division of the aryepiglottic folds and resection of the arytenoid mucosa regardless of the anatomic site responsible for the laryngomalacia. Zalzal et al described using scissors to trim the aryepiglottic folds, lateral edges of the epiglottis, and the mucosa over the arytenoids and corniculate cartilages. Similarly, Marcus et al reported using scissors to trim the obstructing mucosa from the lateral edge of the epiglottis, aryepiglottic folds, and arytenoid cartilage in all patients, even though they saw two distinct patterns of laryngomalacia in their patients; half exhibited anteromedial collapse of aryepiglottic folds and cuneiform cartilage, while the other half manifested only anteromedial collapse of the mucosa overlying the arytenoids. Jani et al performed a suction test similar to the one described by Polonovski et al to assess how much redundant arytenoid mucosa to resect, but unlike Polonovski et al, they always excised the aryepiglottic fold. Roger et al began by sectioning only the aryepiglottic folds, but they later also resected the redundant arytenoid mucosa, occasionally with the corniculate cartilage; of 115 cases, 4 required an epiglottopexy for significant epiglottic wavying. Finally, Kelly and Gray used the laser to vaporize the cuneiform cartilage and an adjacent wedge of the aryepiglottic fold, but they did so only unilaterally; only 17% of their patients required the same contralateral procedure in 2 to 3 months.

We have taken an approach that is similar to Kelly and Gray's by performing supraglottic redundant mucosa resections unilaterally. In only 1 of our patients did we find that a bilateral resection was required, and even this proved to be insufficient as the patient ultimately required a tracheostomy. It is important to note that this patient was 1 of the 2 patients who had been born prematurely with bronchopulmonary dysplasia; supraglottoplasty was successful in all 8 of our full-term infants.

There are authors who advocate tailoring the supraglottoplasty to the structural etiology of the laryngomalacia without relying on a classification system. Zeitouni and Marcus favor the cuneiform cartilage and an adjacent wedge of the aryepiglottic fold, but they did so only unilaterally; only 17% of their patients required the same contralateral procedure in 2 to 3 months. Similarly, Marcus et al performed a suction test similar to the one described by Polonovski et al to assess how much redundant arytenoid mucosa to resect, but unlike Polonovski et al, they always excised the aryepiglottic fold. Roger et al began by sectioning only the aryepiglottic folds, but they later also resected the redundant arytenoid mucosa, occasionally with the corniculate cartilage; of 115 cases, 4 required an epiglottopexy for significant epiglottic wavying. Finally, Kelly and Gray used the laser to vaporize the cuneiform cartilage and an adjacent wedge of the aryepiglottic fold, but they did so only unilaterally; only 17% of their patients required the same contralateral procedure in 2 to 3 months.

In conclusion, it is possible, of course, to perform selected procedures in accordance with an algorithm that does not involve a classification system. However, we find that a classification system focuses the surgeon's attention preoperatively and intraoperatively to the particular offending anatomy, thereby avoiding the use of a shotgun surgical approach.

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