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Laryngomalacia and Its Treatment

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Objective: **To determine 1) airway outcome of infants with laryngomalacia who do not undergo routine direct laryngoscopy (DL) and bronchoscopy (B), 2) the age at resolution of laryngomalacia, and, 3) outcome of supraglottoplasty as a function of the type of laryngomalacia and the presence of concomitant disease.** ***Study Design:*** **Retrospective chart review.** ***Methods:*** **The records of all infants diagnosed with laryngomalacia by flexible fiberoptic laryngoscopy (FFL) between 1990 and 1998 in the Department of Otolaryngology—Head and Neck Surgery, University of Iowa (Iowa City, IA) were reviewed. The type of laryngomalacia was designated by a new classification scheme (types 1–3) based on the site of supraglottic obstruction and the type of supraglottoplasty indicated, should the patient later require surgical intervention. The log rank test was used to compare age at resolution and outcome between types of laryngomalacia and between infants with isolated laryngomalacia versus those with additional congenital abnormalities and/or severe neurological compromise.** ***Results:*** **The type of laryngomalacia was evident in 48 of the 58 charts reviewed and included type 1 (57%), type 2 (15%), type 3 (13%), or combined types (15%). Twenty percent had severe neurological compromise and/or multiple congenital anomalies. The median time to resolution of stridor in these patients was not significantly delayed when compared with infants who had isolated airway anomalies (36 and 72 wk, respectively, vs. 36 wk for isolated laryngomalacia; $P < .4$). Time to resolution did not correlate with the type of laryngomalacia. In 22 infants, clinical symptoms or findings suggested a synchronous airway lesion, and direct laryngoscopy and bronchoscopy were performed. In 11 infants, a second airway lesion was diagnosed (in four cases by FFL and in 7 cases by direct laryngoscopy and bronchoscopy). Complications did not arise in infants who did not undergo direct laryngoscopy and bronchoscopy. Eleven infants with severe laryngomalacia required surgical intervention. The success of supraglottoplasty did not correlate with the type of laryngomalacia or the presence of other congenital anomalies.** ***Conclusions:*** **Routine direct laryngoscopy and bronchoscopy as part of the evaluation of laryngomalacia are not warranted. Performing these procedures should be based on clinical and physical evidence of a concomitant airway lesion. In general, laryngomalacia will resolve within the first year of life, even in children with multiple congenital anomalies and/or severe neurological compromise. The proposed classification scheme is advantagous in that it is simple and correlates the site of obstruction with the surgical procedure most likely to effect a cure, should the patient require a supraglottoplasty. Surgical management is necessary in approximately 15% to 20% of affected infants.** ***Key Words:*** **Congenital laryngeal stridor, laryngeal anomalies, laryngomalacia, supraglottoplasty.**

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INTRODUCTION

Laryngomalacia is the most common cause of stridor in infants. It accounts for 65% to 75% of all cases of stridor[1,2] and is caused by the collapse of supraglottic structures during inspiration (Fig. 1). A self-limited condition, it usually resolves without therapy by 12 to 18 months of age[3]; however in 10% of affected persons, the upper airway obstruction is severe enough to cause apnea or failure to thrive, necessitating surgical intervention. Usually, supraglottoplasty resolves the obstruction; tracheotomy, once the mainstay of surgical therapy, is rarely required.[3,4]

The etiology of laryngomalacia remains unknown. Because of the high incidence of neuromuscular problems in affected infants, some investigators[5] believe that the disorder represents a form of laryngeal "hypotonia." Other investigators suspect that gastroesophageal reflux disease (GERD), which is found in 35%[6] to 68%[7] of affected infants, may play a role, perhaps by inducing supraglottic edema and changing airway resistance enough to potentiate airway obstruction. However, it is also possible that laryngomalacia causes GERD by altering the normal intraabdominal/intrathoracic pressure gradient that typically protects against reflux events.

Laryngomalacia may occur in isolation or in association with other anomalies of the airway or other organ systems. Synchronous airway lesions are reported in up to 19% of affected infants, and for this reason, some investigators mandate direct laryngoscopy and bronchoscopy (DL

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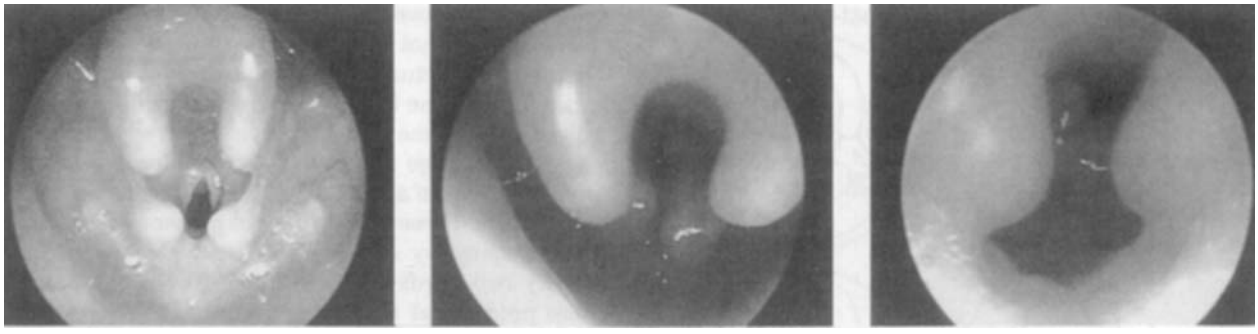

Fig. 1. Laryngoscopic examination of an infant with laryngomalacia. Note patent airway during expiration (**left**) and gradual worsening of obstruction of supraglottis during middle and end inspiration (**middle** and **right,** respectively).

and B) in all infants with laryngomalacia to avoid the risk of missing a potentially life-threatening synchronous airway lesion.[8] However, this recommendation remains controversial,[9] with many surgeons basing the decision to perform DL and B on clinical history, since significant, life-threatening synchronous airway lesions are rare.[9]

During the past 8 years, we have performed DL and B only in select infants with laryngomalacia who presented with apnea, failure to thrive, or features inconsistent with isolated laryngomalacia. This study reviews the outcome in our patient population. Information regarding median time to resolution of stridor, whether isolated laryngomalacia resolves sooner than nonisolated laryngomalacia, and whether the site of supraglottic prolapse affects the time to resolution are reported. This information will aid in the counseling of parents and caretakers of infants with laryngomalacia. A new classification scheme for laryngomalacia is proposed based on the site of prolapsing supraglottic structures and the method of supraglottoplasty used to manage the patients requiring surgical intervention. This classification scheme was implemented after the first 4 years of the study were initiated.

MATERIALS AND METHODS

The charts of all infants diagnosed with a laryngeal anomaly from 1990 to 1998 were retrospectively reviewed to identify the infants with laryngomalacia. Flexible fiberoptic laryngoscopy (FFL) was performed in all infants. Demographic characteristics, complications during gestation, presence of other congenital anomalies, presence of gastroesophageal reflux (GER) or GERD, and symptoms of airway obstruction (including apnea and failure to thrive) were recorded.

Findings at the time of FFL were reviewed. Laryngomalacia was classified into the following types to reflect the tissue causing the supraglottic obstruction: type 1, prolapse of the mucosa overlying the arytenoid cartilages; type 2, foreshortened aryepiglottic folds; and type 3, posterior displacement of the epiglottis (Fig. 2). This classification scheme was devised and implemented during the latter 4 years of the series to aid in determining the appropriate supraglottoplasty procedure for infants who subsequently required surgical intervention. Laryngomalacia in infants enrolled in this series during the first 4 years was classified retrospectively by this scheme when possible by the drawings and narrative description of the site of supraglottic obstruction as determined by FFL. Some infants had combined collapse, usually types 1 and 2. Synchronous airway lesions diagnosed by FFL or by DL and B were noted, as were indications for supraglottoplasty or tracheotomy.

The operative procedures in the small subset of patients with laryngomalacia who required intervention were reviewed. In patients undergoing supraglottoplasty, the technique employed was based on the type of laryngomalacia that was present (Fig. 3). Surgical correction of type 1 laryngomalacia involved excision of redundant mucosal tissue on the posterolateral aspect of the arytenoids using either cold steel dissection or the CO_2 laser. Type

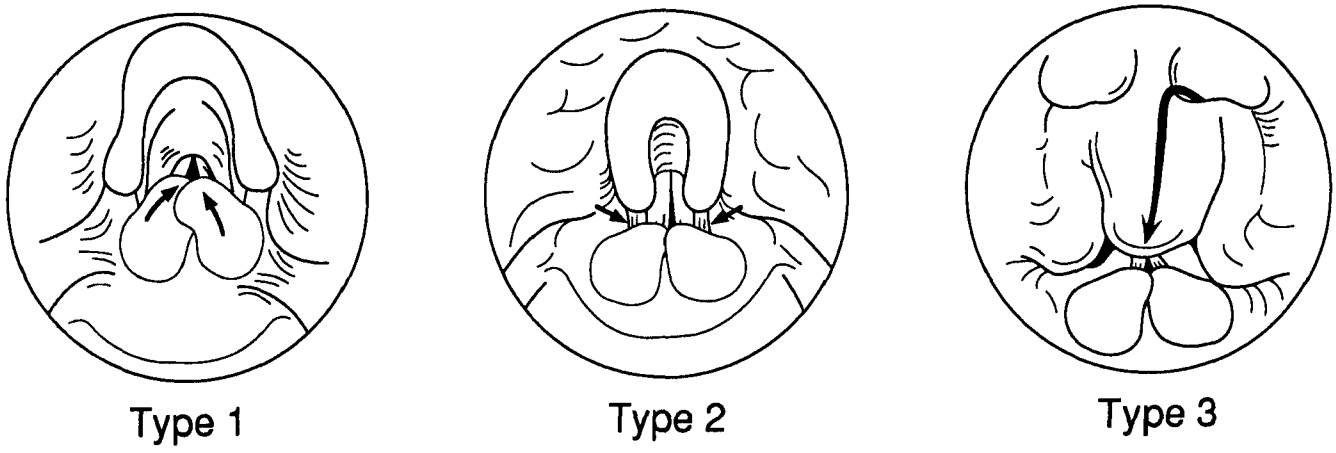

Fig. 2. Classification of laryngomalacia based on site of supraglottic obstruction. Type 1, prolapse of mucosa overlying the arytenoid cartilages; type 2, foreshortened aryepiglottic folds; type 3, posterior displacement of the epiglottis.

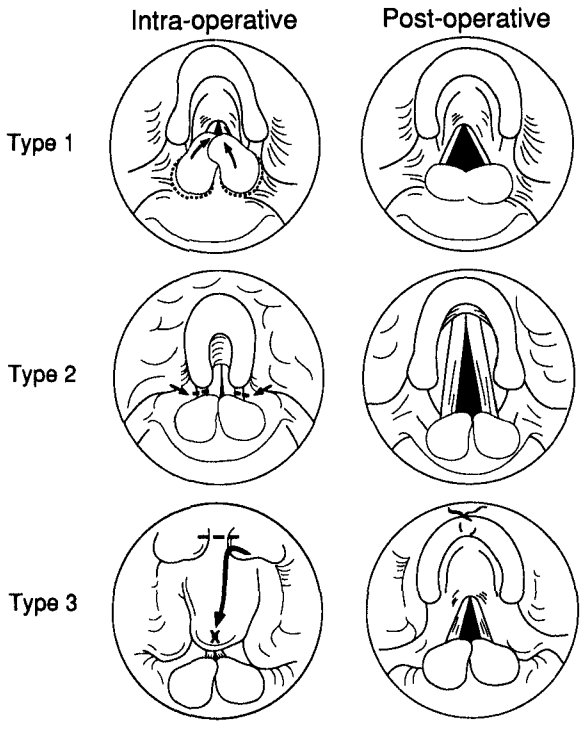

Fig. 3. Type of laryngomalacia predicts the type of supraglottoplasty performed in infants requiring surgical intervention. Arrows depict source of supraglottic obstruction. Surgical management of type 1 laryngomalacia involves excision of mucosa overlying the arytenoid cartilages (dashed lines) while preserving the interarytenoid region. Surgical management of type 2 laryngomalacia involves excising the short aryepiglottic folds (dashed lines) that tether the supraglottic structures in close anterior and posterior approximation. Surgical management of type 3 involves division of the median glossoepiglottic ligament (dashed lines) and suture suspension of the epiglottis to the base of tongue.

2 laryngomalacia was corrected by dividing the short aryepiglottic folds tethering the anterior and posterior supraglottic structures in close approximation. Type 3 laryngomalacia was treated by excision through the glossoepiglottic ligament to retract the epiglottis anteriorly and by placement of a suture from a small denuded region of the epiglottis to a small denuded region of the base of the tongue.

Outcome was reviewed with respect to age at resolution of stridor (that age when stridor was absent or present only with *severe* agitation). Age at resolution and necessity of surgical correction were correlated with the type of laryngomalacia (types 1–3), presence of other congenital anomalies, and/or presence of neurological compromise using the log rank test. Kaplan-Meier curves were generated to compare age at resolution for the different groups of infants. The success of surgical correction was reviewed, as was the airway outcome in infants who did not undergo DL and B.

RESULTS

Fifty-eight infants (white, 54; black, 2; Hispanic, 2) with laryngomalacia were evaluated in the Division of Pediatric Otolaryngology during the study period. All infants underwent FFL, at which time the diagnosis of laryngomalacia was established. Typically, each infant was reevaluated 2 to 3 months after the initial diagnosis and then every 4 to 6 months until symptoms resolved or markedly improved. Ten infants with very mild laryngomalacia were not followed on a routine basis but were instructed to return if symptoms did not improve in several months. One infant's laryngomalacia remained unresolved at the time of this study.

The average age at onset of stridor was 2.2 weeks, and the average age at diagnosis in our department was 3.8 months. Three infants did not present with stridor but with respiratory distress or apnea shortly after birth. Nearly two-thirds (64%) of infants were boys, and most were products of term pregnancies (12% were born preterm, 26–35 wk). Nearly 41% were products of complicated pregnancies (Table I).

Isolated laryngomalacia was present in 35 infants. Congenital anomalies not affecting the airway were present in 27 infants; 7 had multiple anomalies (Table II). Neurological compromise was present in 5 and synchronous airway lesions were detected in 11 of the infants. More than half of the infants (53%) had clinical or subjective evidence of excessive "spitting up" as determined by caretaker history. Of 17 infants (29%) who underwent a diagnostic study for GERD (barium esophagogram or extended pH probe monitoring), 8 were diagnosed with pathological GER.

The type of laryngomalacia (site of supraglottic collapse) was determined from reviewing the charts containing the new classification scheme, diagrams, and/or a narrative description of the findings noted at FFL. The site or sites of supraglottic collapse were able to be determined in 47 of 58 patients as follows: type 1 laryngomalacia, 57%; type 2 laryngomalacia, 15%; and type 3 laryngomalacia, 12%. Seven additional patients (15%) had combined supraglottic collapse, usually types 1 and 2. Most infants (84%) had self-limited laryngomalacia, with mild symptoms not requiring surgical treatment.

The median time to resolution for isolated laryngomalacia was 36 weeks, and by 72 weeks, 75% of infants were free of stridor, even during periods of agitation (Fig. 4). The median time to resolution did not vary significantly in infants with severe neurological compromise (36 wk, $P < .4$) or in infants with other congenital anomalies (72 wk, $P < .4$) (Fig. 4). Seventy-five percent resolution of disease was seen in these latter groups by 44 and 82 weeks, respectively.

TABLE I.
Occurrence of Complicated Pregnancies.

Pregnancy Complication	No.
Maternal tobacco abuse	5
Preterm labor	3
Maternal kidney stones	2
Preeclampsia	2
Maternal diabetes	1
Maternal multiple sclerosis	1
Polyhydramnios	1
Maternal ethanol abuse	1
Vaginal infection	1
Gestational anemia	1
Uncomplicated pregnancy	40

TABLE II.
Incidence of Other Congenital Anomalies.

Congenital Disorder	No.
Ventricular septal defect	3
Cutaneous hemangioma	3
Generalized hypotonia	3
Micrognathia	3
Cleft palate	2
Patent ductus arteriosus	2
Hypertelorism	2
Beckman–Weidman syndrome	1
Down's syndrome	1
CHARGE syndrome	1
Cat-eye syndrome	1
Caroli's disease	1
Stickler syndrome	1
Status epilepticus	1
Congenital glaucoma	1
Cardiomegaly	1

The median time to resolution did not vary with the type of laryngomalacia and was 36 weeks for type 1 laryngomalacia, 72 weeks for type 2 laryngomalacia, 40 weeks for type 3 laryngomalacia, and 26 weeks for combination disease ($P < .4$) (Fig. 5). Seventy-five percent resolution occurred at 44, 72, 40, and 56 weeks, respectively ($P < .4$).

Nine infants (16%) required surgical intervention for laryngomalacia attributable to episodes of apnea[5] and/or failure to thrive.[6] A supraglottoplasty tailored to manage the type of laryngomalacia as designated by the site of supraglottic collapse was performed in each case; one infant required a revision supraglottoplasty to remove residual redundant tissue. The overall success of supraglottoplasty was 78%, since two patients required tracheotomy for continued severe symptoms. One of these two infants also had severe GERD and required a fundoplication.

Two of 27 infants with type 1 disease, 3 of 7 with type 2 disease, 1 of 6 with type 3 disease, and 1 of 7 with combined disease required surgery. The number of surgical procedures performed for each type of laryngomalacia was too small for statistical analysis. Of the two patients undergoing tracheotomy, one had type 1 and the other had type 3 laryngomalacia.

All infants underwent a thorough otolaryngological examination including FFL. Twenty-two infants also underwent DL and B to exclude the presence of a synchronous airway lesion, to further evaluate a laryngeal lesion diagnosed by FFL, or to exclude other airway anomalies before performing supraglottoplasty. Fourteen of these infants were scheduled for DL and B at the time of the initial evaluation, and eight were scheduled for the procedures when follow-up evaluation revealed worsening or changing symptoms. Concomitant airway lesions were found in 11 of these 14 infants, or 19% of the total study population. Four of these lesions were diagnosed by FFL; the remaining lesions were diagnosed by DL and B.

Of the synchronous airway lesions found in 11 infants, surgical intervention was performed in 2 infants, or 4% of the total study population. One lesion that was found via FFL, a cyst of the posterior glottis, was excised. One other infant with tracheomalacia, subglottic stenosis, and paralysis of the left vocal cord underwent laryngotracheal reconstruction. The lesions in the remaining infants did not require treatment. All infants who did not undergo routine DL and B were followed and gradually improved without the need for additional intervention.

DISCUSSION

Our study population is indicative of a tertiary-care referral center with a relatively high percentage of complicated pregnancies. The male predominance we found also was noted by Holinger.[1] Many patients had other congenital anomalies, but none of the anomalies were preferentially associated with laryngomalacia. Type I laryngomalacia was most common. It should be noted that, while other classification schemes for laryngomalacia have been proposed,[1] our scheme is advantageous in that it is simple and correlates the site of obstruction with the surgical procedure most likely to affect a cure, should the patient require a supraglottoplasty. It is important to determine the type of laryngomalacia by FFL preoperatively, since findings during DL and B may vary because of the effect of general anesthesia and placement of a laryngoscope.

Laryngomalacia in isolation is a benign process that resolves spontaneously in 70% of infants by 1 year of age. Improvement occurs gradually, with stridor first becoming intermittent and then occurring only with marked agitation. Although infants with laryngomalacia often have concomitant neurological disorders[5] and/or congenital anomalies, the presence of these conditions does not alter significantly the time to resolution of symptoms, nor does the type of laryngomalacia. This information will aid in counseling parents and caretakers of infants with laryngomalacia.

All infants with stridor should undergo a thorough otolaryngological examination that includes FFL. This study not only establishes the diagnosis of laryngomalacia, but also excludes other anomalies of the larynx that cause stridor. Associated laryngeal disorders evident by FFL can include laryngeal cysts, vocal cord paralysis, vascular malformations, neoplasms, subglottic hemangiomas, paradoxical vocal cord motion, posterior glottic stenosis, and glottic webs. If isolated laryngomalacia is documented by FFL, careful attention must be made to verify that the endoscopic findings correlate with the infant's respiratory phase of stridor and with the severity of the infant's presenting symptoms.

Although debate exists concerning whether all infants with laryngomalacia should undergo DL and B to detect synchronous airway lesions, we perform these procedures only in select infants. Included in this category are the following:

1. Infants with laryngomalacia and severe respiratory distress, failure to thrive, apnea events, or recurrent pneumonia.

2. Infants with symptoms that do not match the degree of laryngomalacia noted by FFL.

3. Infants with synchronous lesions of the larynx.

4. Infants who are likely to require supraglottoplasty.

Of the 22 infants who underwent DL and B in this series, 7 had other airway lesions that were not diagnosed by FFL but only 1 required an additional surgical procedure (laryngotracheal reconstruction) directed at this anomaly (Table III). In total, 2 of 58 infants (3.4%) required surgical intervention for a synchronous airway lesion.

The parents or caretakers of all infants with laryngomalacia in this series were counseled about the natural history of the disease and instructed to contact us if additional symptoms appeared, if new concerns arose, or if the frequency or severity of the stridor worsened. Eight of the 22 patients undergoing DL and B were initially observed and later underwent endoscopy to evaluate worsening symptoms; one of these patients was found to have mild tracheal compression by the innominate artery that did not require additional intervention.

Following these guidelines, all clinically significant synchronous airway lesions requiring intervention were detected, and 36 of 58 patients avoided unnecessary DL and B. This study confirms the findings of Mancuso et al.[9] that synchronous airway lesions requiring therapy are uncommon in infants with laryngomalacia (3.4%) and, when they do occur, undoubtedly present with obvious signs and symptoms that alert the astute otolaryngologist to the necessity of DL and B.

Nine of 58 patients required surgical management of the laryngomalacia because of its severity. The number of patients was too small to determine whether the need for surgery or its success correlated with the type of laryngo-

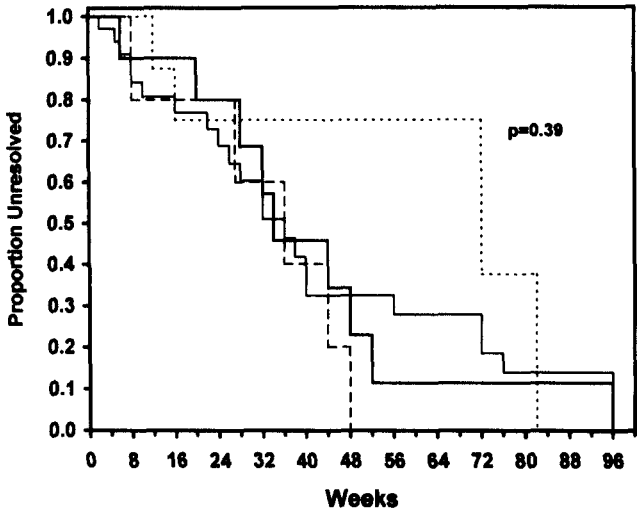

Fig. 4. Kaplan-Meier curve depicting age-at-resolution (in weeks) in infants grouped by type of laryngomalacia. Infants with multiple congenital anomalies with isolated and nonisolated laryngomalacia (dotted curve) require more time to resolution. Patients with severe neurologic compromise (dashed curve) and synchronous airway lesions (bold solid curve) behaved similarly to infants with isolated laryngomalacia (solid curve).

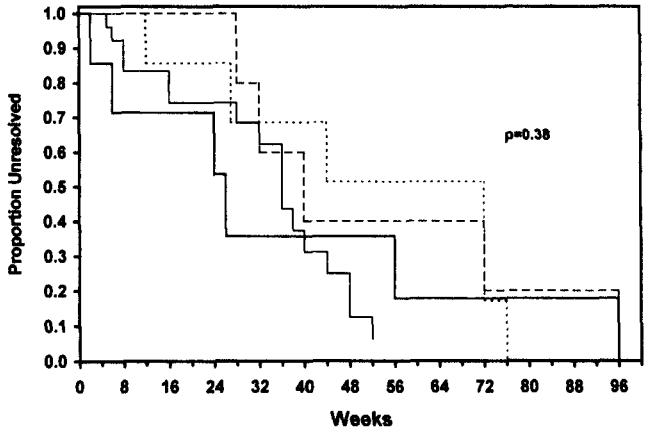

Fig. 5. Kaplan-Meier curve demonstrating age at resolution (in weeks) versus type of laryngomalacia (type 1 = solid curve; type 2 = dotted curve; type 3 = dashed curve; combined supraglottic collapse = bold solid curve). No significant difference was noted by site of supraglottic collapse.

TABLE III.
Synchronous Airway Lesions.

Patient No.	Symptom(s)	Age at Evaluation	Synchronous Airway Lesion(s)	Method of Diagnosis	Treatment
1	Dysphagia	2 wk	Type 1 laryngeal cleft	DL and B	None
2	FTT/apnea/cyanosis	3 wk	Mucus retention cyst on glottis	FFL	Cyst excision
3	Cyanosis/apnea	2 mo	Innominate artery compression	DL and B	None
4	Chronic cough	8 mo	Tracheomalacia	DL and B	None
5	Hypoxia/bradycardia/FTT	6 mo	Mild subglottic stenosis	DL and B	None
6	FTT/cyanosis	1 mo	Tracheomalacia/subglottic stenosis	DL and B	None
7	Hypoxia/bradycardia/FTT	2 mo	Tracheomalacia/subglottic stenosis/VC paralysis	DL and B	Tracheotomy
8	None	3 days	Left VC paralysis	FFL	None
9	None	3 mo	Grade I subgottic stenosis	FFL	None
10	None	2 days	Grade I subgottic stenosis	FFL	None
11	FTT/apnea with feeding	12 mo	Aberrant subclavian artery	DL and B	None

FTT = failure to thrive; DL = direct laryngoscopy; B = bronchoscopy; FFC = flexible fiberoptic laryngoscopy; VC = vocal cord.

malacia or with the presence of other non–airway-related anomalies.

Tracheotomy was necessary in two infants. One of these infants failed two supraglottoplasty procedures and later underwent surgical correction of severe GER, which may have contributed to the failed supraglottoplasties. The other infant had tracheomalacia, subglottic stenosis, and paralysis of the left vocal cord, which were believed to contribute too much to the infant's symptom manifestation to warrant a supraglottoplasty.

CONCLUSION

In the majority of infants, laryngomalacia follows a benign course, resolving by 1 year of age. It is often accompanied by concomitant disorders that can confound and confuse the clinical picture, but usually does not alter the disease duration significantly. The role of GERD may be significant; however, its contribution to the pathogenesis and course of disease is not well understood at present. The type of laryngomalacia does not significantly alter the disease course, nor does it appear to predict the success of surgical correction.

Routine performance of DL and B is not indicated in all patients with laryngomalacia. If infants with laryngomalacia continue to have severe symptoms that cannot be attributed to laryngomalacia or if the severity of symptoms increases, a thorough examination of the upper aerodigestive tract is warranted. Supraglottoplasty may be necessary in approximately 16% of infants with the disease and is very effective in lessening or eliminating the obstruction. Tracheotomy is rarely necessary to manage patients with laryngomalacia.

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