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Research Article

# Laryngomalacia: The Importance of Disease Severity

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## Abstract

**Introduction:** Assessment of Laryngomalacia (LM) disease severity is a complex task. To date, there are severity scoring systems that use subjective findings to stratify disease. We propose to create an objective scoring system based on an objective scale, to improve disease stratification, treatment and follow up.

**Objectives:** 1. Illustrate the importance of LM disease severity. 2. Propose an objective scoring system to standardize LM management.

**Method:** We performed a literature review of the most common symptoms of Laryngomalacia. We assigned adjusted points to those symptoms based on their severity as follows: Inspiratory stridor-1, retractions-1, choking/gagging-1, difficulty feeding-1, failure to thrive-2, apnea-3, and pectus excavatum-3. Disease severity was stratified as follows: mild 1-3, moderate 4-6, severe>6. A retrospective analysis of 182 LM charts at a single tertiary care center and we applied our scoring system to first encounter. We then investigated for a correlation between markers of disease severity such as reflux therapy, incidence of surgery and higher scores.

**Results:** In our overall cohort, 148 patients had mild disease, 24 moderate, and 10 severe. Reflux therapy ($p<0.008$) and incidence of surgery ($p<0.001$) both significantly correlated with higher scores. The Chi-square analysis of a score of 4 was the highest (25.3) and significance was attained with $p<0.001$. 15 patients underwent supraglottoplasty: 8-mild, 4-moderate, 3-severe. Two thirds of patients classified as severe underwent tracheostomy. All patients were on Ranitidine.

**Conclusion:** We successfully showed that our scoring system correlated with disease severity. Also, we suggest a cut-off score of 4 to decide for surgery. Our scoring simplifies disease severity categorization and should be validated in a larger multi-center prospective study.

**Keywords:** Laryngomalacia, Score, Severity Level of Evidence: 4

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## Introduction

Although Laryngomalacia is the most common cause of infantile inspiratory stridor, determining and assessing its severity is a very complex task. The hallmark symptom is inspiratory stridor that worsens with feeding, excitement, agitation, crying and supine positioning [1]. The symptoms begin at birth or within the first few weeks of life, peak at 6 to 8 months, and typically resolve by 12 to 24 months [2].

The characteristic inspiratory stridor is secondary to the supraglottic collapse of the airway. The common findings seen on exam that illustrate the disease are the "prolapse of the posteriorly positioned arytenoid cartilages and mucosa into the airway during inspiration, shortening of the distance between the arytenoid and epiglottis, and an "omega-shaped" or "retroflexed epiglottis" [2,3]. All or just one of these characteristics in the supraglottis may be observed on direct visualization. Olney et al. created a scoring system of the type of laryngomalacia based on the site of supraglottic collapse. Type 1 consists of arytenoid cartilage prolapse, type 2 consists of shortened Aryepiglottic (AE) folds, and type 3 consists of epiglottis collapse [4].

Diagnosis is highly suspected by clinical history and confirmed by direct visualization. Direct visualization is gold standard for diagnosis. This is achieved by flexible laryngoscopy in office, or Direct Laryngoscopy and Bronchoscopy (DLB) in the operating room.

The most common comorbidity associated with laryngomalacia is acid reflux occurring in 65% to 100% of patients [2]. The nature of the relationship between acid reflux and laryngomalacia has been investigated without any conclusive evidence suggesting that it contributes to the etiology of laryngomalacia [5]. Several papers showed that the supraglottic collapse of the airway creates a negative intrathoracic pressure which promotes acid reflux onto the laryngeal tissues causing laryngeal edema [4-6]. Literature is clear on the fact that disease severity is correlated with incidence of reflux symptoms [7]. The second most common finding associated with laryngomalacia is a Secondary Airway Lesion (SAL), whose incidence is recorded as 7.5%-64% [1]. The presence of a SAL may worsen the disease process at times [1].

Management of laryngomalacia is determined by clinical judgment of progression of disease and its severity. The severity of disease is characterized by the gravity of the symptoms. It is categorized into mild, moderate or severe. Mild laryngomalacia usually presents with inspiratory stridor and hardly any feeding difficulties. Moderate to severe laryngomalacia presents with failure to thrive, apnea/ALTE (Apparent Life-Threatening Event), choking, gagging, and/or retractions [2,4]. Although several attempts have been made to produce an

objective classification system, most centers continue to classify disease based on clinical symptoms and judgment [2,4].

Many patients with mild laryngomalacia can be treated symptomatically whereas patients with mild-moderate disease may need symptomatic treatment and surgical intervention at times [1,2,5]. Patients with severe laryngomalacia may require a surgical intervention such, as a supraglottoplasty or epiglottopexy [1,4,5]. To date there is no objective quantitative measure assessing the severity of Laryngomalacia on which to base treatment.

In our literature search, there have been attempts to create an algorithm to the approach of treatment of a child with laryngomalacia based on general characterizations of severity of disease [1]. By introducing a scoring system to the severity of disease, we hope to provide a more quantitative algorithm to aid physicians in the treatment approach for laryngomalacia patients.

The scope of disease and management of laryngomalacia has varied in the literature. Primary objective of this study is geared towards developing an algorithm for the treatment of laryngomalacia after reviewing the disease process of 274 patients at our institution. Our algorithm will be based on a disease severity scoring system based on presenting symptoms. Based on this new scoring system, we will aim to provide guidelines on the surgical management of these patients.

## Materials and Methods

This retrospective review was conducted at Children's Hospital of Michigan (CHM) following approval by Wayne State University Investigational Review Board. The clinical database of the Pediatric Ear Nose Throat (ENT) Associates practice at CHM was used to generate a list of patients from 2004 to 2011 with the ICD-9 code of 748.3 (Congenital Anomalies of larynx trachea and bronchus). The following data were collected on a data collection sheet from each chart by an approved reviewer. Age at presentation, duration of symptoms until ENT visit, gestational age, postnatal complications, presenting symptoms, diagnostic procedure, and findings at diagnosis, treatment, and age at resolution of disease were recorded from each chart. There were two reviewers blinded to each other and the patients. Reviewers were asked to select "yes" or "no" with each corresponding data. No scoring system was used in data collection to eliminate bias from reviewers.

Inclusion criteria included: Age 0-3 years, complete follow up throughout the duration of the disease, diagnosis of disease by visualization only. Exclusion criteria included: Age > 3 years, incomplete follow up, and co-morbidities that contribute to airway compromise (craniofacial anomalies or underlying neurologic anomalies).

A scoring system was created to classify the disease severity of laryngomalacia based on the presenting symptoms. Symptoms that are common and in a milder form were assigned a score of 1. These symptoms include inspiratory stridor, retractions, gagging/choking, and difficulty feeding (defined as insufficient duration of feeding or inability to breathe appropriately while feeding). More severe symptoms were assigned higher scores and those include apnea/ALTE (Apparent Life-Threatening Event - Assigned a score of 3), Failure to Thrive (FTT - Assigned a score of 2), and pectus excavatum (Assigned a score of 3). Failure to thrive was defined as BMI or weight less than the 3rd percentile. The individual symptom scores were added up to a total. The maximum total score attainable in the presence of all symptoms is 12. The total scores were then categorized as mild (score 1-3), moderate (4-5), or severe (6 or greater).

The means of visually confirming diagnosis consisted of flexible fiberoptic in the office or direct visualization in the OR. If no direct visualization was performed, patient was excluded.

There were a total of five surgeons whose patients were reviewed. Patient confidentiality was protected by assigning a letter to each patients' initials included in the study which was entered in a master list on a locked computer.

Since this is a retrospective chart review, the symptom severity score of those who have received a surgical intervention will be compared to those who did not receive a surgical intervention. The long-term plan of this research is to perform a prospective confirmatory analysis of the disease severity score identified in this research.

Statistics: Initial analyses in this project include performing an independent t-test on the two independent groups (surgical intervention vs. no surgical intervention) disease severity score. It is hypothesized that children who received a surgical intervention will have a significantly higher disease severity score. We have a priori hypothesized that this score will be above 4 for those who received surgical intervention and below 4 for those who did not. Thus, a Pearson's chi-square test was performed for the different cutoff scores. We report the analysis of a 2 x 2 contingency table: surgical intervention Yes/No vs. a disease severity score ≤ or > 4.

## Results

Of the 274 patients with code of laryngomalacia, 182 had complete charts that could be reviewed. Among the 182 patients, mean gestational age was 35.0 weeks (SD = 3.3, range = 24 - 39); 77.9% were full-term. There were more males (62.8%) in the sample than females (37.2%). The modal racial group was Caucasian patients (42.1%). Table 1 summarizes the demographics of the patient population. Among the neonates, 13.1% required a NICU stay and 10 (5.7%) were intubated/ventilated.

| Number of Patients (Valid Percent) | |
|---|---|
| **Race** | |
| Black | 61 (35.7%) |
| White | 72 (42.1%) |
| Hispanic | 4 (2.3%) |
| Asian | 2 (1.2%) |
| Middle Eastern | 1 (0.6%) |
| Indian | 1 (0.6%) |
| Other | 13 (7.6%) |
| Unknown | 17 (9.9%) |
| Missing data | 11 |
| **Gender** | |
| Female | 113 (62.8%) |
| Male | 67 (36.8%) |
| Missing data | 2 |
| **Term at Birth** | |
| Term | 134 (77.9%) |
| Pre-term | 38 (22.1%) |
| Missing data | 10 |
| **Total** | 182 |

**Table 1:** Demographics.

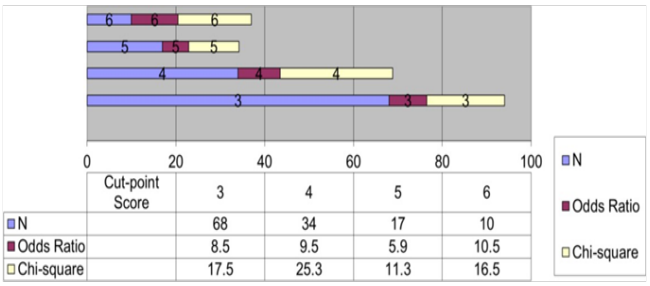
On average, reported symptoms lasted for 17.8 weeks (SD = 20.2, range = 0.4 – 156.0 weeks). Laryngomalacia diagnosis was made using a flexible fiberoptic scope in the office or DLB in the operating room. Flexible fiberoptic diagnosis was made in 43% of the patients and 41% diagnoses were made in the operating room. The remaining patients had a magnified airway as their initial diagnostic procedure, but still subsequently undewerwent flexible laryngoscopy. The anatomic abnormalities identified during exam included Omega-shaped epiglottis (49%; N = 88), shortened aryepiglottic folds (56%; N = 101), arytenoid prolapse into airway (58%; N = 105), and supraglottic edema (40%; N = 73). Just over one fourth of the sample (27%; N = 48) had a secondary airway lesion (tracheomalacia, bronchomalacia, subglottic stenosis, or vocal fold paralysis).

All symptoms at presenting visit were recorded. All 182 patients had inspiratory stridor on presentation. Table 2 lists the presenting symptoms from most frequent to least frequent. Most of the initial visits took place in the office (N = 144). There were 22 inpatient consultations in which the diagnosis was made, 8 ER consultations, and 4 patients which were diagnosed after an intraoperative consultation with a diagnostic DLB.

| | Frequency of symptoms (Percentage) |
|---|---|
| **Stridor (1 point)** | 182 (100%) |
| Choking/Coughing (1 point) | 85 (47%) |
| Retractions (1 point) | 66 (37%) |
| Difficulty feeding (1 point) | 20 (20%) |
| Apnea (3 point) | 22 (12%) |
| Failure to thrive (2 point) | 9 (5%) |
| Pectus excavatum (3 point) | 0 (0%) |

**Table 2:** Symptoms.

Reflux therapy ($p < 0.008$) and surgery ($p < 0.001$) were highly correlated with higher disease severity scores. Chi-square analysis and odds ratio were calculated for each score of 3, 4, 5, and 6. The Chi-square analysis of a score of 4 was the highest at 25.3, and significance was attained with $p < 0.001$. The odds ratio of a score of 4 was 9.6 with a 95% confidence interval ranging from 3.5 to 26. Tables 3a and 3b show the 2 x 2 contingency tables for observed and expected results. Figure 1 shows the chi square analysis and odds ratio of the subsequent scores. To further evaluate score cut-point, an ROC analysis was performed (Figure 2). Using a cut-point of 4 had the highest level of specificity without losing too much sensitivity.




| Cut-point Score | 3 | 4 | 5 | 6 |
|---|---|---|---|---|
| N | 68 | 34 | 17 | 10 |
| Odds Ratio | 8.5 | 9.5 | 5.9 | 10.5 |
| Chi-square | 17.5 | 25.3 | 11.3 | 16.5 |

**Figure 1:** Cut-point Score Analysis.

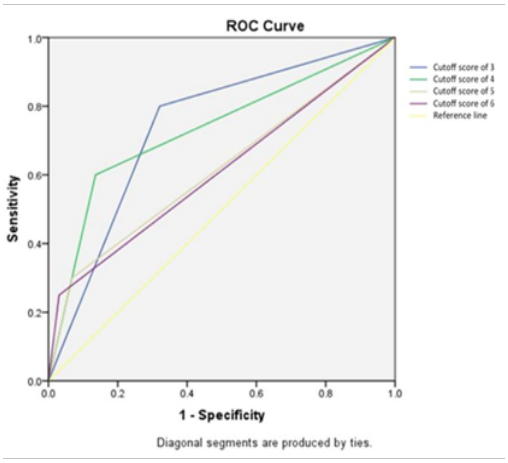



**Figure 2:** ROC Curve for different cut-off scores.

## Discussion

### Laryngomalacia: Demographics and symptoms

Laryngomalacia was found to be more common in males, with a male to female ratio of about 1.7:1. The literature mentions a disease predilection to males with a male to female ratio of 1.5:1 [8,9]. We found that laryngomalacia was more common in Caucasians, in agreement with other sources [8]. Our results concerning the patients' demographics are in concordance with the literature. Similar to other studies, we found laryngomalacia to be more prevalent in term infants [10,11]. It's important to note that we did not perform any racial demographics analysis of the hospital's infant population to rule out potential bias, nor did we assess the rate of seeking of medical treatment for the different races involved (Table 3a).

| | Surgery | No Surgery | Total |
|---|---|---|---|
| Score > 4 | 12 | 22 | 34 |
| Score < 4 | 8 | 140 | 148 |
| Total | 20 | 162 | 182 |

**Table 3a:** Two by two table showing observed results for a cutoff score of 4.

Symptoms of laryngomalacia are numerous the most common of which is inspiratory stridor [2,8,12]. All our patients had inspiratory stridor on presentation. Feeding problems, which include choking/coughing, are the second most common set of symptoms according to the literature [2,8,10,12]. Indeed the second most common symptom in our sample was choking/coughing happening at a rate of 47% (Table 3b).

| | Surgery | No Surgery | Total |
|---|---|---|---|
| Score > 4 | 4 | 30 | 34 |
| Score < 4 | 16 | 132 | 148 |
| Total | 20 | 162 | 182 |

**Table 3b:** Two by two table showing expected results for a cutoff score of 4.

Reflux symptoms are commonly associated with laryngomalacia, with 90.1% of our patients suffering from them, in concordance with the rate of 65-100% mentioned in the literature [2]. Secondary airway

lesion is also prevalent co-morbidity associated with laryngomalacia occurring at a rate of 28.7% in our sample. Sources in the literature approximate the rate of secondary airway lesions to be ranging from 7.5-64% [1,12].

### Laryngomalacia: Attempt at classification

Laryngomalacia is a burdensome disease that significantly decrease patients' quality of life [13,14]. Its presentation and severity however vary from a self-limited condition to a life- threatening one requiring surgical intervention. In effect, most infants suffering from laryngomalacia are only followed by their primary physician and are never referred to an otolaryngologist [8]. There is however around 10-20% cases severe enough to be referred to a specialist for potential surgical management [5,12]. Since Laryngomalacia has such a wide spectrum of disease, many have suggested ways to classify it [8,15,16]. A popular classification is that developed by Thompson et al., and it is mainly based on subjective criteria. It classifies cases into mild ones if only inspiratory stridor is present, with resting $spO_2$ greater than 98%; moderate ones if inspiratory stridor is also associated with frequent feeding problems and choking and a $spO_2$ of 95-96%; severe cases if symptoms also include apnea, cyanosis, failure to thrive and a resting $spO_2$ of 85-86%. According to that classification 40% of cases are mild, 40% are moderate and 20% are severe [1,2,8,12]. The objective of this paper is to suggest a more objective score based on the symptoms of laryngomalacia along with a cutoff score that will facilitate the decision to proceed with surgery. The purpose of this score is not to abide by it blindly but rather to use it as a supporting algorithm in decision-making. According to our classification, our sample consists of 148 mild cases, 24 moderate cases, and 10 severe cases. The score correlated with disease severity as evidenced by the high correlation between severity indicators (i.e. reflux ($p < 0.008$) and incidence of surgery ($p < 0.001$)) and higher scores. We found that a cut-off score of 4 best predicted surgical procedure. Our recommendation is to highly consider surgery whenever score exceeds 4. The decision to perform surgery has to however been corroborated by clinical judgment.

The retrospective nature of this paper leads to some discrepancies in our results. Indeed, of the 20 patients who proceeded to surgery, 8 were classified as mild. This discrepancy can be explained by the fact that those patients were only scored upon presentation and not immediately before surgery. All in all, we think that this score should be validated by a prospective multicenter study.

## Conclusion

In this study we suggest an objective scoring system to stratify laryngomalacia into mild, moderate or severe disease. Our data analysis shows that our higher scores are significantly associated with disease severity. Also, a score higher than 4 is significantly associated with surgical intervention in this group. Hence, the authors suggest using a cutoff score of 4 to decide for surgery. However, because of the retrospective nature of this study some discrepancies were noted, and this scoring system needs to be validated by a multicenter prospective study.

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## Potential Conflicts of Interest

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