Management Outcome of Severe Laryngomalacia at Queen Sirikit National Institute Child Health

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Background: *Outcomes of the different management in severe laryngomalacia (LM) have not been evaluated.*
Objective: *To identify the management practices and to evaluate the outcomes in patient with severe LM.*
Material and Method: *The medical records of LM at Queen Sirikit National Institute Child Health between January2007 and December 2012 were retrospectively reviewed.*
Results: *Severe LM 69.8% (30/43) were found in patients diagnosed with LM. Type B (complete collapse) at 46.67% were the most common finding. Decision of management were made individually based on consideration of disease severity and comorbidity. The outcomes after management were evaluated by pre- and post-symptoms score. Post-symptoms scores were statistically significant better than pre-symptom score in all management (observation $p<0.001$, laser supraglottoplasty $p = 0.003$, and tracheotomy $p = 0.001$).*
Conclusion: *Our management in severe LM include: observation, laser supraglottoplasty, and tracheostomy. The overall post-management outcome were satisfactory but the present study was limited to relatively small number of patients.*

Keywords: *Laryngomalacia (LM), pre- and post-symptoms score, Tracheostomy, Laser supraglottoplasty*

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Laryngomalacia (LM) is considered one of the most common cause of early onset of infantile stridor resulted in upper airway obstruction due to the collapse of supraglottis during inspiratory phase[1]. Definite diagnosis can be achieved by fiberoptic laryngoscopy. Level of severity such as mild or moderate usually resolved by the age of 12 to 24 months[1]. Severe cases required surgical intervention. The initial process generally requires either tracheostomy[1,2,3,6] or supraglottoplasty, which is currently the preferred approach[1,4,5-8].

Objective

The present study aimed to identify the management practices and to evaluate their outcomes in severe LM patients.

Material and Method

We conducted a retrospective study among pediatric patients with LM receiving care at Queen Sirikit National Institute Child Health (Children's Hospital, Bangkok) between 2007 and 2012. All cases were diagnosed by direct visualization of larynx by fiberoptic laryngoscopy done by an otolaryngologist. This method was used to confirm the diagnosis by determining the dynamic movement of the laryngeal structure during spontaneous ventilation under general anesthesia using Sevoflurane or Propofol with 10% Xylocaine spray without endotracheal intubation[2,6].

The disease was classified into three level of severity[8], mild cases were those presented with inspiratory stridor with no other symptoms, moderate cases were those with stridor, cough, chocking, regurgitation, and feeding difficulty, without hypoxia or desaturation, and severe cases were presented with stridor, aspiration, feeding difficulty, pectus excavatum, cyanosis, apneic pause, failure to thrive, and aspiration[1,8]. The classification of LM was made in accordance with the location of the supraglottic structure collapse from the classification system developed by Holinger and Konior[3], type A, posterolaterally located (short aryepiglottic fold and redundant supra-arytenoid soft tissue), type B, completely located (long tubular curled epiglottis), and type C, anteriorly located (posterior displacement of epiglottis to posterior pharyngeal wall).

The pre- and post-symptoms scores were measured by applying Lee et al's method i.e., the presence of each symptom was scored as 1 and the absence as 0 (zero) score[9]: inspiratory stridor, suprasternal retraction, substernal retraction, feeding

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difficulty, chocking, post-feeding vomiting, failure to thrive, and cyanosis. If there were two symptoms the score is 2 and so on with the highest score as 8.

The comorbidity conditions included neurological disorder, epilepsy, Down syndrome, heart disease, gastroesophageal reflux disease, and past intubation.

Management and surgical procedures

Information regarding the onset of the symptom, the age at diagnosis using fiberoptic laryngoscopy, gestational age, pre- and post-management symptoms scores, comorbidities, type of LM, disease severity, length of hospital stay, and treatment cost were obtained from medical records abstraction.

The treatment decisions were based on shared and informed decision making, taking into consideration disease severity, and comorbidity. For non-severe LM cases (mild-moderate level), observation management were generally employed. For severe cases, having respiratory compromise or feeding difficulty with three or more comorbidities including one of neurological disease, Down syndromes, complex heart disease, or past intubation, then tracheostomy was done[1,2,6]. Patient with less than three comorbidities, supraglottoplasty was done[1]. Patient with no comorbidity and with corrected morbidity were managed by observation.

The surgical decision was based on the indication as recommended by Richter and Thompson[1]. CO_2 laser system was mounted on an operating microlaryngoscope. The CO_2 laser supraglottoplasty procedure was modified according to the location of the laryngeal prolapsed. Tracheostomy was done as conducted according to Casselbrant[11].

Post-operative care and follow-up

In the laser group, all cases were observed in the recovery room until patient's conditions was stable. The patients' respiration and oxygen saturation were closely monitored during the first 24 hours post-operation. Antibiotics were administered for one week. Steroid and antireflux medication were not routinely administered. In general, the patients were allowed to resume oral intake within two hours and kept under observation overnight and discharged the following day.

For those who underwent tracheostomy, standard post-operative care for tracheostomy wound and tube were followed. Antibiotics were administered for an average of one week duration.

All cases were followed for two months post-operative for the evaluation of post-management symptoms scores.

Statistical analysis

The baseline data on clinical characteristics and outcomes were descriptively analyzed according to gender, gestational age, the onset of the symptoms, the age at diagnosis, medical comorbidities, types (A, B, or C), severity (mild, moderate, or severe) of LM, management options such as observation, laser supraglottoplasty, tracheostomy, length of stay, and cost of treatment.

Wilcoxon statistic test a non-parametric analysis was used to compare the pre- and post-management symptoms score in severe LM, this was due to non-normal distribution of data. Statistical analysis was performed by using SPSS software version 15.

Results

The patients' basic demographic information was presented in Table 1. The LM patients were composed of 22 males and 21 females. The median onset of the symptoms usually occurred one day of infant age (IQ1:IQ3 1:14) and the diagnosis of the

Table 1. Demographic data and clinical features of the patients (n = 43)

Variable	Median (Q1:Q3) or number (%)
Onset of symptoms (days)	1 (1:14)
Age at diagnosis (months)	2.47 (1.33:4.63)
Gender	
Male	22 (51.16)
Female	21 (48.84)
Gestational age	
Term	30 (69.76)
Preterm	13 (30.23)
Severity of laryngomalacia	
Mild	5 (11.6)
Moderate	8 (18.6)
Severe	30 (69.8)
Length of stay (days) (n = 25)*	
Tracheostomy	40 (16.75:55.25)
Laser	8 (4:12)
Cost of treatment (baht/patient) (n = 25)*	
Tracheostomy	133,685 (58,377.50:229,403.75)
Laser	37,634 (13,286.0:66,884.0)

* Inpatient

disease by the doctor were made at 2.47 months of infant age (IQ1:IQ3 1.33:4.63). In regard to gestational age, 30 patients at term and 13 patients were preterm. Type A (posterolateral) was the most common among the three types. The length of hospitalization and cost of treatment were higher among the patients receiving tracheostomy than laser treatment (Table 2). Regarding to the degree of disease, 69.8% (n = 30) were in the "severe" group while 18.6% (n = 8) and 11.6% (n = 5) were found to be in the "moderate" and "mild" group, respectively. Patients with the most "severe" LM received all types of management such as observation, laser supraglottoplasty, and tracheostomy. Type B (complete) was the most common in severe LM patients, 46.67% (n = 14). Comorbidity group in type B patients were managed with all treatment such as observation 20% (n = 1), laser supraglottoplasty 18.18% (n = 2), and tracheostomy 42.86% (n = 6). Type B with non-comorbidity group were managed by observation 60% (n = 3) and laser supraglottoplasty 18.8% (n = 2), as seen in Table 3. We found that post-symptoms scores were significantly better than pre-symptom score in management, observation $p<0.001$, laser supraglottoplasty $p = 0.003$, and tracheotomy $p = 0.001$ as shown in Table 4. After laser supraglottoplasty, the three most common symptoms were stridor 41.67% (n = 5/12), substernal retraction 40.0% (n = 4/12), and suprasternal retraction 20% (n = 2/10). The most common symptoms that were managed (100%) were feeding difficulty and chocking. Post-tracheostomy, symptoms of upper airway obstruction and feeding difficulty improved 100% except failure to thrive, which remained at 40%, and post-observation with only substernal retraction remained at 33.3% (n = 1/3).

Table 2. Number and percentage of the patients by severity and management procedure operated

Procedure	Severity, n (%)		
	Mild	Moderate	Severe
Observation	5 (100)	8 (100)	5 (16.67)
Laser supraglottoplasty	0	0	11 (36.67)
Tracheostomy	0	0	14 (46.67)
Total	5 (100)	8 (100)	30 (100)

Discussion

Among our clinical samples, the onset and age at diagnosis were congruent with the existing literature on LM[1] i.e., symptoms begin at birth or within the first few weeks of life and LM was usually diagnosed within the first four months of life. In regard to disease severity, we identified 11.6% and 18.6% were classified as "mild" and "moderate" respectively, which was lower compared to 40% of each level as reported by Thompson[8]. It showed that 69.8% of our samples had severe LM, as compared to Thomson, which had 20%[8]. This could be due to referral bias as

Table 3. Number and percentage of severe laryngomalacia (LM) patients by types of LM, medical comorbidity, and treatment procedure (n = 30)

Type of LM		Treatment procedure, n (%)		
		Observation (n = 5)	Laser supraglottoplasty (n = 11)	Tracheostomy (n = 14)
Type A (n = 4)	Non-comorbidity (n = 2)	1 (20.00)	1 (9.09)	0
	Comorbidity (n = 2)	0	0	2 (14.29)
Type B (n = 14)	Non-comorbidity (n = 5)	3 (60.00)	2 (18.18)	0
	Comorbidity (n = 9)	1 (20.00)	2 (18.18)	6 (42.86)
Type C (n = 12)	Non-comorbidity (n = 1)	0	1 (9.09)	0
	Comorbidity (n = 11)	0	5 (45.45)	6 (42.86)

Table 4. Comparison of the average pre- and post-treatment symptom scores of severe LM patients by treatment procedure

Treatment		Median (Q1:Q3)	*p*-value
Observation (n = 5)	Pre-symptoms score	4.0 (3.5:5)	<0.001
	Post-symptoms score	0 (0:1)	
Laser supraglottoplasty (n = 11)	Pre-symptoms score	4.0 (3:5)	0.003
	Post-symptoms score	1.0 (0:2)	
Tracheostomy (n = 14)	Pre-symptoms score	4.5 (2.75:6.25)	0.001
	Post-symptoms score	0 (0:0.25)	

p-value at 95% for Wilcoxon statistics test

we are a tertiary center for children in central region of Thailand. The reason for doing laser treatment first was that laser supraglottoplasty was very popular and the success rate was 90%, with very low complication rate (less than 5%)[10]. Furthermore, the cost was not high compared to tracheostomy. In the present study, including the admission cost, the median hospital charge at our center for tracheostomy was 133,685 Thai Baht per patient, which was higher than that of laser supraglottoplasty at 37,634 Thai Baht per patient. In addition, length of stay among those receiving laser surgery were shorter than those who had tracheostomy. Therefore, it is essential to take into consideration underlying comorbidity of individual patients when deciding on appropriate treatment option.

As compared to Holinger and Konior (1989)[3], the most common type was type A (posterolateral collapse). The present report showed that type B (complete collapse) 46.67% were the most common in severe LM.

In our report, all cases of LM with muscular hypotonia associated with neurological disorder and/or Down syndrome underwent tracheostomy. For other underlying comorbidities, the decision should be made individually. As suggested by Toynton et al, endoscopic aryepiglottoplasty is the operation of choice for severe LM, however, underlying neurological disease reduced the success rate of this intervention[12]. Similarly, patients with neurological and cardiac comorbidities generally required tracheostomy at significantly higher rate compared to their counterpart[10].

Overall symptoms significantly improved in 82.6% of patients after laser supraglottoplasty as reported by Lee et al (2007)[9]. In our report, we found out that all post-management scores, included laser treatment, were improved significantly.

In regard to the improvement of symptoms after laser supraglottoplasty in severe LM, Lee et al[9] indicated that the substernal and suprasternal retraction were most likely to be improved with this procedure while the two least improved were feeding difficulty and choking. In contrast to our study, choking and feeding difficulties were the most improved 100% while the two least improved were stridor (58.33%) and substernal retraction (60%). This finding suggests that it is usual for symptoms to persist for a few months after intervention. Therefore, it is essential for future study that the follow-up period be extended to at least six months[8] to assess the resolution of symptoms and other complications that may occur later.

Conclusion

The overall post-management outcome for severe LM were satisfactory. However, the study was limited because of the relatively small number of patients.

What is already known on this topic?

Historically, the surgical management of LM was tracheostomy associated with high morbidity. In the last 20 years, supraglottoplasty, an alternative to tracheostomy, has been a successful and safe surgical endoscopic approach. In severe LM patients with medical comorbidities, tracheostomy is the choice for management after report of failure using laser. There is no report regarding the outcome of the following three management, laser supraglottoplasty, tracheostomy, and observation.

What this study adds?

The present study supports the literature of the improved outcome after supraglottoplasty. The other managements, tracheostomy and observation also had good outcome. The decision for management depends on the patients status.

Potential conflicts of interest

None.

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การดูแลรักษาผู้ป่วยโรคกล่องเสียงอ่อนระดับรุนแรงในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี

นาฎยพร จรัญเรืองธีรกุล

ภูมิหลัง: *โรคกล่องเสียงอ่อนระดับรุนแรงมีการดูแลรักษาหลายวิธีซึ่งยังไม่พบรายงานการประเมินผลหลังการดูแลรักษาแต่ละวิธีในประเทศไทย*

วัตถุประสงค์: *เพื่อศึกษาในรายละเอียดของแต่ละวิธีการรักษาในโรคกล่องเสียงอ่อนระดับรุนแรงและประเมินผลหลังการรักษา*

วัสดุและวิธีการ: *เป็นการศึกษาย้อนหลังจากข้อมูลที่บันทึกจากผู้ป่วยที่วินิจฉัยโรคกล่องเสียงอ่อนระดับรุนแรงในสถาบันสุขภาพเด็กแห่งชาติมหาราชินี ตั้งแต่ เดือนมกราคม พ.ศ. 2550 ถึง ธันวาคม พ.ศ. 2552*

ผลการศึกษา: *ผู้ป่วยโรคกล่องเสียงอ่อนระดับรุนแรงพบ 69.8% จากผู้ป่วยโรคโรคกล่องเสียงอ่อนทั้งหมด ชนิด บี เป็นชนิดที่มีการยุบตัวลงของอวัยวะเหนือกล่องเสียงทุกทิศทางซึ่งพบมากที่สุด การตัดสินใจรักษาแต่ละวิธีขึ้นอยู่กับผู้ป่วยแต่ละรายรวมทั้งปัจจัยของโรค และโรคประจำตัว ประเมินผลการรักษาโดยใช้คะแนนรวมของอาการก่อนและหลังรักษา พบว่าคะแนนรวมหลังการรักษาดีกว่าก่อนรักษาทุกวิธีอย่างมีนัยสำคัญทางสถิติ ได้แก่ การสังเกตติดตาม $p<0.001$ เลเซอร์อวัยวะเหนือกล่องเสียง $p = 0.003$ และเจาะคอ $p = 0.001$*

สรุป: *พบว่าผู้ป่วยโรคกล่องเสียงอ่อนระดับรุนแรงมีผลหลังการรักษาดีขึ้นทุกวิธี แต่จำกัดด้วยผู้ป่วยมีจำนวนน้อย*
