Endoscopic supraglottoplasty in children with severe laryngomalacia with and without neurological impairment

José C. Fraga, Luciano Schopf, Vanessa Volker, Simone Canani

Abstract

Objective: to describe indications and results of supraglottoplasty for severe laryngomalacia in children with or without neurological impairment.

Methods: eight children with severe laryngomalacia submitted to endoscopic supraglottoplasty were retrospectively studied. Four had neurological impairment (male, mean age 6 years), and 4 did not present neurological problems (3 female, mean age 11.5 months). Surgery indications were respiratory distress, feeding difficulties, failure to thrive, and low oxygen saturation. Polysomnographic evaluation was carried out on the last 2 children, showing abnormal oxygen saturation, obstructive apnea, and hypoventilation. All children received preoperative antibiotics and corticosteroids.

Results: all children without neurological impairment had significant relief of symptoms. Children with neurological impairment had different outcome: one needed tracheotomy immediately after surgery due to edema and supraglottic granulation tissue. The other three children presented initial relief of symptoms, but subsequent follow-up showed progressive airway obstruction: one needed another endoscopic surgery 6 months later; other needed tracheotomy 7 months later. The children who were not submitted to tracheostomy presented persistent severe airway obstruction. No endoscopic surgery complication was observed.

Conclusions: 1) Endoscopic supraglottoplasty is well tolerated and does not present complications when used in children; 2) Endoscopic supraglottoplasty was efficient in the treatment of children with severe laryngomalacia and in without neurological impairment; however, supraglottoplasty did not resolve airway obstruction in children with neurological impairment.


Introduction

Laryngomalacia is a disorder characterized by collapse of laryngeal cartilage during inspiration with glottic obstruction. It is the most common disorder of the larynx, and it is the most frequent cause of stridor in children. Studies have indicated that laryngomalacia relates to 50 to 75% of diagnoses in patients with laryngeal malformation.1,2

The characteristic manifestation of laryngomalacia is inspiratory stridor. Stridor is usually manifest in the first
two weeks of life and deteriorates with crying, agitation or exercise, but improves with neck extension. The final diagnosis of laryngomalacia is obtained with endoscopy of the respiratory tract on patients breathing spontaneously. The exam can indicate presence and severity of obstruction caused by collapse of laryngeal cartilage.

Most children with laryngomalacia present a favorable prognosis since the ventilatory obstruction is not intense and there is significant improvement of symptoms before two years of age. However, in approximately 10% of cases the ventilatory obstruction presents severe and can cause several complications, including life-threatening apnea, cyanosis, respiratory failure, feeding difficulties, failure to thrive, and cor pulmonale. Until recently, tracheostomy was indicated for laryngomalacia patients; however, due to complications of tracheostomy in children, endoscopic excision of redundant supraglottic tissue (supraglottoplasty, epiglottoplasty, or aryepiglottoplasty) became the first option for treatment of children with severe laryngomalacia.

Certain children with cerebral palsy develop stridorous breathing much similar to that in laryngomalacia of infants. Endoscopic findings in pediatric patients have shown presence of redundant arytenoid mucosa, small aryepiglottic ligament, and floppy epiglottis also much similar to that in laryngomalacia. This type of laryngomalacia is called neurasthenic laryngomalacia and is probably a result of a disorder in innervation of pharynx and larynx muscles. Good results have been reported with endoscopic surgery in neurasthenic laryngomalacia patients.

It is our objective to retrospectively assess the results of endoscopic supraglottoplasty (SGP) in children with and without cerebral palsy who presented severe laryngomalacia.

**Patients and methods**

We carried out a retrospective study on medical records of eight pediatric patients submitted to endoscopic supraglottoplasty for severe laryngomalacia at the Hospital Moinhos de Vento and the Hospital de Clínicas de Porto Alegre, both located in the city of Porto Alegre, southern Brazil, from January, 1995 to February, 2000. Out of the eight patients, four had cerebral palsy and four did not present neurologic disease (Table 1). Age average of patients with cerebral palsy was six years, for a median of eight years and seven months; in the group without cerebral palsy, the age average was 11.5 months, for a median of 10.5 months. Patients were considered candidates for endoscopic surgery when presenting resting or strain respiratory difficulties, swallowing difficulties, failure to thrive, and low transcutaneous oxygen saturation. Respiratory difficulties were considered severe when patients presented intercostal or sternal recession, cyanosis, and feeding difficulties with frequent choking and failure to thrive. The last two patients to be operated on were also submitted to polysonography, which indicated several desaturation episodes during oxymetry and apnea and severe hypoventilation during sleep. Depending on suspected diagnosis, children were also submitted to chest X-ray, gastroesophageal reflux study, and echocardiogram.

Laryngomalacia was diagnosed by bronchoscopy on patients submitted to general anesthesia and breathing spontaneously. Even after the diagnosis of laryngomalacia was established, complete endoscopic examination of the airways was carried out to eliminate related malformations. During surgery, patients were submitted to laryngoscopy for exposure of the larynx and microsurgery instruments were introduced (scissors and tongs) into the mouth to section and remove redundant supraglottic tissue or lateral edge of the epiglottis (Figure 1). Endoscopic surgery varied according to the type of obstruction of each patient (Figure 1). All patients were administered corticoid (dexamethazone) and prophylactic antibiotics (cephalothin) preoperatively for 48 hours.

If possible, children were not intubated during or after surgical procedures. Patients were admitted to the Pediatric Intensive Care Unit during the postoperative for observation, and were administered nebulized adrenaline in case of any ventilatory difficulty.

**Results**

All patients in the study population presented stridor and severe breathing difficulties that were the cause of several hospitalizations and emergency care situations.

All patients with neurological disease were boys and presented cerebral palsy (Table 1). One of these patients presented Cornelia de Lange syndrome and two others gastroesophageal reflux controlled with medication.

![Figure 1 - Types of endoscopic surgery in children with severe laryngomalacia: A) Excision of redundant arytenoid mucosa; B) Section of aryepiglottic fold; C) Removal of the lateral edge of the epiglottis](image-url)
Endoscopic findings indicated bilateral arytenoid (100%) and epiglottis (25%) collapse; indications for surgery were related to resting respiratory difficulties (75%) and low transcutaneous oxygen saturation (50%).

In the case of patients without neurological disease, there were three girls and one boy (Table 1). None of these patients presented concomitant diseases. Endoscopic findings were the same as those for neurological disease patients; indications for surgery were related to resting and/or strain respiratory difficulties (75%), failure to thrive (50%), and swallowing difficulties (25%). In addition, the last two children to be operated on presented abnormal polysomnographic study.

The type of endoscopic surgery was the same for both groups (Table 2). Patients with neurological disease required tracheal intubation soon after the surgical procedure. One patient presented significant improvement of ventilatory dysfunction after surgery with occasional episodes of mild stridor; however, stridor and ventilatory dysfunction deteriorated progressively and tracheostomy was necessary seven months after endoscopy. Another patient presented subglottal edema and supraglottal granulation tissue, hindering extubation and leading to tracheostomy six days after endoscopic surgery. Yet another patient presented initial improvement after surgery; however, this patient presented gradual recurrence of respiratory obstruction and another endoscopic surgery was required six months later. In reoperation, the redundant tissue of the right-side arytenoid was reresected. Initially, this patient presented good evolution but later follow-up indicated that the child persisted with stridor and intermittent respiratory dysfunction. Finally, the last patient with cerebral palsy to be operated on presented initial improvement of ventilatory obstruction but persisted with mild, intermittent stridor with significant deterioration of the obstruction leading to several hospitalizations and emergency care situations due to severe respiratory dysfunction. The average follow-up of these patients was two years and one month.

The evolution of patients with severe laryngomalacia but no neurological disease is presented in Table 2. There was no need for tracheal intubation after surgery despite patients being frequently submitted to nebulized adrenaline during the immediate postoperative period due to ventilatory dysfunction. This condition deteriorated soon after surgery. All patients presented complete improvement of ventilatory obstruction despite mild, intermittent stridor in most patients. There was also a significant decrease in hospitalizations and emergency care situations due to acute ventilatory dysfunction. The average follow-up of these patients was one year and 11 months.

**Discussion**

Laryngomalacia is usually manifest a few days after birth with respiratory stridor. In children with cerebral palsy, stridor starts at a later stage in life, usually late childhood. This characteristic explains the difference in age average of our population in relation to laryngomalacia with or without neurological disease.

| Table 1 - Characteristics of children with severe laryngomalacia with or without neurological disease, submitted to endoscopic supraglottoplasty |
|---|---|---|---|
| **Sex** | Neurological disease (n=4) | Absence of neurological disease (n=4) |
| **Age - surgery** | Male - 4 | Male - 1; Female - 3 |
| – mean (variation) | 6 years (5 to 1 mo –13 yrs) | 11.5 months (1 mo – 1 to 8 mo) |
| – median | 8 years 7 months | 10.5 months |
| **Associated malformation** | GER – 2 | None |
| | Cornelia de Lange syndrome – 1 | |
| **Indication for surgery** | Resting respiratory difficulties – 3 | Resting or strain respiratory difficulties – 4 |
| | Abnormal oximetery – 2 | Failure to thrive – 2 |
| | | Abnormal polysomnography* – 2 |
| | | Swallowing difficulty – 1 |
| **Endoscopic findings** | Arytenoid collapse bilateral – 4 | Arytenoid collapse bilateral – 4 |
| | Epiglottis collapse – 1 | Epiglottis collapse – 1 |

* Abnormal oximetery, severe apnea, and hypoventilation during sleep.
Table 2 - Type and result of endoscopic surgery in children with severe laryngomalacia, with and without neurological disease

<table>
<thead>
<tr>
<th>Type of surgery</th>
<th>Neurological disease (n=4)</th>
<th>Absence of neurological disease (n=4)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td>- Unilateral arytenoid resection - 2</td>
<td>- Unilateral arytenoid resection - 2</td>
</tr>
<tr>
<td></td>
<td>- Bilateral arytenoid resection - 2</td>
<td>- Bilateral arytenoid resection - 2</td>
</tr>
<tr>
<td></td>
<td>- Partial epiglottectomy - 1</td>
<td>- Partial epiglottectomy - 1</td>
</tr>
<tr>
<td>Need for intubation after surgery</td>
<td>- All patients (mean of 3.2 days)</td>
<td>- None</td>
</tr>
<tr>
<td>Initial results</td>
<td>- Initial improvement of ventilatory obstruction - 3</td>
<td>- Complete improvement of ventilatory obstruction - 4</td>
</tr>
<tr>
<td></td>
<td>- Intermittent stridor - 3</td>
<td>- Intermittent stridor - 3</td>
</tr>
<tr>
<td></td>
<td>- Hindered extubation (tracheostomy) - 1</td>
<td></td>
</tr>
<tr>
<td>Need for new surgery</td>
<td>- New arytenoid resection unilateral (after 6 months) - 1</td>
<td>- None</td>
</tr>
<tr>
<td>Follow-up (average time)</td>
<td>- Progressive deterioration of ventilatory obstruction (one tracheostomy after 7 months) - 3</td>
<td>- Intermittent stridor - 3</td>
</tr>
<tr>
<td></td>
<td>- Maintenance of previous tracheostomy (2 years 1 month) - 1</td>
<td>(1 year 11 months)</td>
</tr>
</tbody>
</table>

The identification of children with associated malformations is an important stage in the treatment of severe laryngomalacia. Cases of tracheal or bronchial alterations should be excluded by endoscopy considering that abnormalities in more distal areas of the airways usually need to be treated first. This is an important consideration at the moment of endoscopy, since full examination of the airways should be carried out even after the diagnosis of severe laryngomalacia is established. Diagnosis and treatment of concomitant gastroesophageal reflux is also important in children with severe laryngomalacia, since this condition can deteriorate respiratory difficulties caused by malacia. In this sense, before classification of the severity of laryngomalacia, it is necessary to control cases of gastroesophageal reflux.7

Most children with laryngomalacia do not require any surgical intervention since flaccidity of supraglottic cartilage improves with age, with symptoms disappearing completely up to two years of age.1,2 This is also the case for children with cerebral palsy and laryngomalacia, considering that only a minority of these patients will develop severe ventilatory obstruction requiring treatment.6 Pediatric patients with severe laryngomalacia, defined as those patients with obstructive dyspnea, cyanosis, failure to thrive, and cor pulmonale, require surgical treatment.5 It is important to underscore that severity of laryngomalacia does not depend on intensity of stridor, but rather on manifestations as a result of ventilatory obstruction.7

Traditionally, children with severe laryngomalacia used to be submitted to tracheostomy. In the past decade, endoscopic excision of the supraglottic tissue has been reported as effective in the treatment of these children. In 1922, Iglauer carried out the first endoscopic surgery for treatment of stridor by performing epiglottectomy.8 In 1928, Hasslinger reported excellent results in resection of aryepiglottic folds of three patients with laryngomalacia.9 In 1944, Schwartz suggested that severe laryngomalacia could be treated by surgical resection of lateral edge of the epiglottis. However, it was not until 1984 that Lane et al.11 proposed for the first time that severe laryngomalacia could be treated by endoscopic resection of the redundant mucosa of one or both arytenoids, and by resection of lateral edge of the epiglottis. Since then, several studies have reported good results with use of this technique.12-15

As we have observed, the most common indication for laryngomalacia surgery is severe ventilatory difficulty due to collapse of supraglottic structures. Others have reported that alterations in polysonographies can be useful for analyzing severity of ventilatory obstruction. According to Zarzal et al.,12 out of a series of 10 operated children, eight presented obstructive sleep apnea. Hollinger and Konior,15 in a report with 13 children, described eight children with sleep apnea and five with repetitive cyanosis. The use of polysonography on the last two patients who were operated on in our population showed that this exam is extremely useful for the indication of surgery, considering that it
quantitates severity of ventilatory obstruction. Consequently, the indication for surgery is no longer based solely on subjective information, but also on numerical results that assess the severity of the obstruction.

Endoscopic excision of redundant supraglottic tissue is a simple technique that was well-tolerated by our patients. Improvement in ventilation was observed soon after the surgical procedures, which can be carried out with surgical instruments or laser. Microsurgical procedures can result in small local bleeding that, in general, ends spontaneously. Despite the fact that laser does not present risk for bleeding, excessive use of laser procedures can result in edema and local granulation tissue with swallowing difficulties or even need for postoperative intubation. Independently of the equipment used, it is important to remove supraglottic tissue as conservatively as possible with minimal excision enough to open the glottis. Depending on the cause of supraglottic ventilatory obstruction, one or more endoscopic surgical procedures can be carried out (removal of one or both redundant arytenoid tissue, sectioning of aryepiglottic ligaments, or removal of lateral edge of the epiglottis - Figure 1). However, it is better to carry out another endoscopic surgery to remove more tissue than to resect in excess on the first intervention and cause complications that are generally difficult to treat.

Results for supraglottoplasty for laryngomalacia were different in children with and without neurological disease. Children without cerebral palsy presented excellent results in early and late follow-up procedures. All of them presented improvement in ventilation despite occasional, mild stridor. Clinical improvement of these children was clearly observable since there was no need for hospital readmission or emergency care due to ventilatory difficulty.

Children with cerebral palsy and severe laryngomalacia presented unsatisfactory endoscopic supraglottoplasty. Two patients required tracheostomy and the other two remained with severe ventilatory dysfunction during long-term follow-up. Our results differ from those of Hiu et al. who described good results with use of the technique in four patients with cerebral palsy and severe laryngomalacia. The authors, however, did not report on duration of follow-up. We observed an early improvement in symptoms but with future recurrence of ventilatory difficulty. This may be a result of development of the neurological disorder of the pharynx and larynx with gradual deterioration of ventilatory dysfunction.

We did not observe complications in endoscopic supraglottoplasty. Complications reported for this procedure are local bleeding, infection and sepsis, synchia of the interarytenoid muscles, supraglottic stenosis, and aspiration.

In conclusion, our study has shown that supraglottoplasty is a safe procedure for pediatric patients causing significant improvement of severe laryngomalacia in children with no neurological disease. Children with cerebral palsy did not present significant improvement following surgery. Further, more extensive studies are needed to establish a final conclusion on usefulness of this surgical procedure in children with severe laryngomalacia.

References


Correspondence:
Dr. José Carlos Fraga
Rua Ramiro Barcelos 2350,sala 600 - 6º andar
CEP 90430-000 – Porto Alegre, RS, Brazil
Phone: +55 51 3316.8232
E-mail: jcfarga@conex.com.br