Journal of Pediatric Surgery 50 (2015) 519–523

Contents lists available at ScienceDirect

Journal of Pediatric Surgery

journal homepage: www.elsevier.com/locate/jpedsurg

Management of laryngomalacia in children with congenital syndrome: The role of supraglottoplasty

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ARTICLE INFO

Article history:
Received 19 March 2014
Received in revised form 21 May 2014
Accepted 24 May 2014

Key words:
Laryngomalacia
Supraglottplasty
Congenital syndrome
Neurologic anomalies

ABSTRACT

Background/Importance: Supraglottoplasty is the surgical procedure of choice for severe laryngomalacia and has shown to be successful in most cases; however, patients with medical comorbidities present a higher rate of failure. To date, the best management of laryngomalacia in children with congenital syndrome remains unclear.
Purpose: To study the outcome of supraglottoplasty in children with severe laryngomalacia, and to analyze the management and outcome in infants with a congenital syndrome.
Methods: Retrospective medical records review from January 2003 to October 2012 of all patients who underwent laser supraglottoplasty for severe laryngomalacia at the University Children's Hospital Zurich, Switzerland.
Results: Thirty-one patients were included; median age at time of surgery was 3.5 months. Three patients (10%) had a genetically proven congenital syndrome with associated neurologic anomalies. Overall success rate was 87%. Failures were observed in four (13%) of 31 cases; including all three patients presenting a congenital syndrome.
Conclusions: Supraglottoplasty is an effective and safe treatment for laryngomalacia in otherwise healthy children. Signs of a possible underlying predominant neurologic origin and discrepancy between the clinical presentation and the endoscopic findings have to be taken into account, as in children with congenital syndrome with neurologic anomalies the risk of failure is higher.

Laryngomalacia is the most common cause of stridor in infants, accounting for up to 75% of all congenital laryngeal anomalies [1]. It is characterized by a dynamic obstruction of the upper airway caused by an inward collapse of supraglottic structures during inspiration. Although several theories have been postulated [2–7], the exact etiology of laryngomalacia is not fully understood and different factors may contribute to the disease [6,8], notably gastroesophageal reflux [9,10]. Laryngomalacia typically presents with a moderate to high-pitched fluttering inspiratory stridor being more marked during increased air demands. The symptoms usually begin within the first weeks of life, progress to a peak around the age of 6 to 9 months and resolve by 12 to 24 months [7,11]. While in most cases treatment consists of watchful waiting, 10%–20% of children with laryngomalacia require further intervention [7,12]. Signs of severity and indications for surgical intervention are dyspnea with suprasternal/intercostal retractions, recurrent cyanosis, hypoxia, life-threatening apneas, feeding difficulties with failure to thrive, cor pulmonale, and right heart failure [7,13]. In these severe forms, an endoscopic examination of the entire upper airway has to be performed to confirm the clinical diagnosis, to characterize the endoscopic findings and to rule out other associated anomalies.

Endoscopic supraglottoplasty is the procedure of choice in case of severe laryngomalacia.

The aims of this study were to: 1) review our patients' outcomes after supraglottoplasty; 2) analyze cases of supraglottoplasty failures in children with congenital syndrome and associated neurologic disease; and 3) identify factors influencing the results.

1. Methods

This retrospective study was conducted at the University Children's Hospital of Zurich, and includes 31 consecutive patients who underwent CO_2 laser supraglottoplasty for severe laryngomalacia from January 2003 to October 2012. Institutional review board approval was obtained for this study.

The diagnosis of laryngomalacia was based on clinical presentation. Severe laryngomalacia needing surgical treatment by supraglottoplasty was defined as severe stridor with dyspnea, usually with suprasternal retractions during inspiration. Other symptoms such as feeding difficulties, failure to thrive, and obstructive apnea or hypoxia were sought but not required for inclusion. These children were considered as requiring endoscopy under general anesthesia for precise upper airway evaluation and therefore no bedside fiberoptic endoscopy was attempted.

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http://dx.doi.org/10.1016/j.jpedsurg.2014.05.035
0022-3468/

The diagnosis of laryngomalacia was confirmed by transnasal fiberoptic laryngoscopy during spontaneous respiration under general anesthesia allowing a dynamic view of the airways including the vocal cord function (Fig. 1). Rigid laryngotracheobronchoscopy was then performed to complete the evaluation of the airways and to rule out synchronous lesions. All endoscopies were video-recorded.

All supraglottoplasties were conducted by the same surgeon under general anesthesia, the patient being ventilated through a small caliber endotracheal tube positioned to minimize interference with the exposure of the concerned supraglottic areas. The suspension laryngoscope was positioned to expose the structures to be resected, and the procedure was performed using the CO_2 laser (Lumenis 30C (Sharplan), superpulse mode, 2.5–3 W) connected to a microscope and a micro-manipulator. The precise surgical technique was adapted to the patient's area of major obstruction (Figs. 2 and 3): section of the shortened aryepiglottic folds and/or resection of the lateral edge of the epiglottis and/or vaporization of redundant mucosa over the arytenoids or epiglottopexy consisting in erecting the epiglottis by creating a raw surface on the base of the tongue and suturing the lingual surface of the epiglottis to the base of the tongue. During the first 5 years of the inclusion period the general attitude in our clinic was to admit the children postoperatively intubated to the intensive care unit and to leave them intubated up to 24 hours. This attitude evolved with growing experience of the different teams involved: extubation was always attempted in the operating theatre and the child admitted to the intensive care unit for overnight monitoring. The records of all patients were reviewed for demographics, preoperative signs and symptoms, comorbidities, endoscopic findings (including evaluation of the video recordings), surgical techniques, postoperative symptoms, complications and outcome. Retrospective analysis of the recorded videoendoscopies was performed. The site of supraglottic obstruction was classified according to Olney et al. [14]: prolapse of the mucosa overlying the arytenoids' cartilage (type 1); shortened aryepiglottic folds commonly associated with a long, tubular, omega-shaped epiglottis that curls on itself (type 2); and overhanging epiglottis that collapses posteriorly (type 3). Furthermore, the severity of the obstruction was graded according to visualization of the vocal cords during respiration: + vocal cords visible during inspiration and expiration, ++ vocal cords visible solely during expiration, and +++ vocal cords nonvisible during inspiration and expiration.

Success of supraglottoplasty was defined as resolution of the initial symptoms without the need for further intervention. Cases with a residual stridor but without labored breathing were also considered as successful. Failure was defined as insufficient improvement of the initial symptoms requiring further intervention such as revision surgery, oxygen therapy, noninvasive ventilation, tube feeding or tracheotomy.

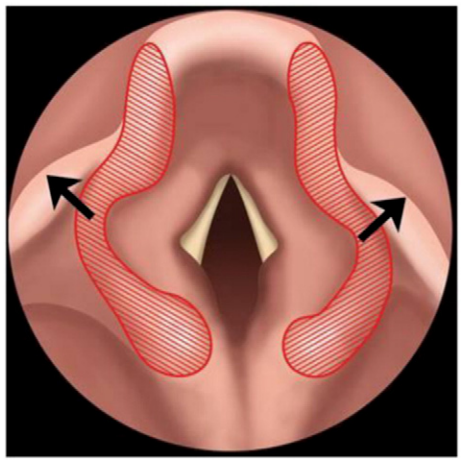

Fig. 2. Method of supraglottoplasty: the precise surgical technique is adapted to the area of major obstruction.

2. Results

Thirty-one patients underwent supraglottoplasty for severe laryngomalacia between January 2003 and October 2012. There were 18 males (58%) and 13 females (42%). The median age at surgery was 3.5 months (1 to 53 months).

Five patients (16%) presented with comorbidities: three (10%) had a genetically proven congenital syndrome with an associated neurologic condition. One child presented with an isolated retrognathia, and one patient had a congenital heart disease (Fallot tetralogy) that had been surgically corrected previously. The median age at surgery of the five children with comorbidities was 4.5 months and 1.5 months for the three children with a congenital syndrome.

Symptoms appeared in 26 cases (84%) during the first 3 weeks of life, and in 14 cases (45%) stridor was noted to be present at birth. At the time of diagnosis all children presented with stridor. In addition, 19 of 31 (61%) children presented with feeding difficulties, 13 (42%) failure to thrive, and 5 (16%) hypoxia and desaturations. No children presented with a pectus excavatus, cor pulmonale or pulmonary hypertension. All three children with a congenital syndrome had

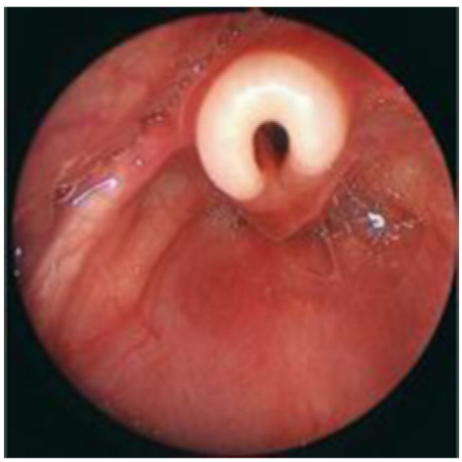

Fig. 1. Endoscopic photograph showing supraglottic airway collapse caused by laryngomalacia.

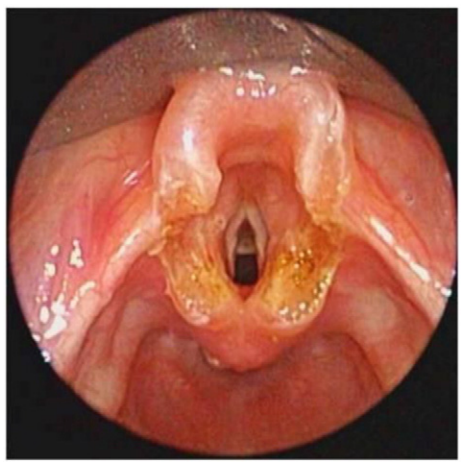

Fig. 3. Endoscopic photograph of the larynx after laser CO_2 supraglottoplasty consisting in section of the shortened aryepiglottic folds, resection of the lateral edges of the epiglottis and vaporization of redundant mucosa over the arytenoids.