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Supraglottoplasty outcomes in children with Down syndrome☆

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ABSTRACT

Objective: Children with Down syndrome have a higher incidence of upper airway obstruction and laryngomalacia. We sought to determine outcomes of supraglottoplasty in this patient population.
Methods: A retrospective chart review was performed from January 2000 through January 2014. Children (n = 18) at our institution with the diagnosis of Down syndrome who underwent supraglottoplasty were included. We reviewed patient characteristics, preoperative findings, and surgical outcomes (stridor, feeding problems, respiratory distress, weight, sleep apnea, and tracheostomy or feeding tube dependence).
Results: The average age at surgery was 7.7 months. Operative indications included feeding difficulties (n = 9), noisy breathing or respiratory distress (or both) (n = 16), and sleep-related symptoms (n = 7). Most patients (89%) were extubated successfully on postoperative day 1. There were 2 major complications (CPAP requirement and aspiration pneumonia) and no perioperative deaths. Fifty percent had improved weight (mean = 18 percentile points). Feedback was available from 88% of parents with 100% reporting improvement in respiratory symptoms and 93% reporting improved feeding. Eight patients (44%) subsequently required either adenoidectomy or adenotonsillectomy. Two patients later underwent tracheostomy, 2 subsequently needed a gastrostomy tube and 2 required revision supraglottoplasty.
Conclusions: The majority of children with Down syndrome and laryngomalacia benefit from supraglottoplasty, with outcomes of improved breathing, feeding, and sleeping.
However, approximately half may require additional airway procedures. This procedure is well tolerated and associated with a low risk of complications especially given their high rate of comorbidities.

1. Introduction

Laryngomalacia is the most common congenital anomaly of the larynx and the top cause of neonatal stridor [1]. Patients usually present within the first 2 weeks of life with inspiratory stridor, although sometimes presentation and diagnosis do not occur until age 3–4 months. The presentation and spectrum of this disorder have been widely characterized, and several theories currently exist to explain the pathophysiology [2].

Generally, cases of laryngomalacia are mild, and approximately 70% of patients will outgrow the phenomenon by age 18–24 months without surgical intervention [2]. In contrast, surgery is warranted if the noisy breathing is not self-limited and leads to episodes of respiratory distress, apnea, cyanosis, pulmonary hypertension, cor pulmonale, or failure to thrive. Approximately 20% of children with laryngomalacia will require surgical intervention, with surgical options depending on endoscopic findings [3]. All procedures are performed endoscopically and are categorized under the umbrella term “supraglottoplasty.”

Laryngomalacia may be an isolated finding or one of several medical comorbidities. The most common and likely universal comorbidity is laryngopharyngeal reflux (from gastroesophageal reflux disease) [3]. Secondary airway lesions such as tracheomalacia and subglottic stenosis affect approximately 50% of patients with laryngomalacia [4]. Neurologic or congenital heart disorders may also be present, complicating both the diagnosis and management of laryngomalacia. Finally, the presence of syndromes and genetic disorders, most commonly Down syndrome (DS), often obscure the treatment of laryngomalacia. Upper airway obstruction is the reason for otolaryngology consultation in 75% of children

Abbreviations: AHI, apnea-hypopnea index; OSA, obstructive sleep apnea; POD, postoperative day; DS, Down syndrome.

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with DS [5]. A recent study by Hamilton et al. estimated the incidence of laryngomalacia in the DS population to be approximately 1% [6]. However, in a small series of children undergoing upper airway endoscopy due to significant respiratory morbidity, 50% of children with Down syndrome were found to have laryngomalacia compared with 19% of children without DS [7].

An abundance of research on the otolaryngologic manifestations of DS has emerged in the past 2 decades. However, little has been reported about the success rate of surgical intervention for patients with Down syndrome and a concurrent diagnosis of laryngomalacia. Here, we describe how supraglottoplasty in these patients affects symptoms of stridor, feeding problems, respiratory distress, cyanotic episodes, weight, and sleep apnea.

2. Methods

We performed a retrospective chart review of select patients seen at Mayo Clinic (Rochester, Minnesota) from January 2000 through January 2014. After receiving study approval from the Mayo Clinic Institutional Review Board, we used a database search system to identify children (ages 0–18 years) with a diagnosis of Down syndrome who underwent supraglottoplasty. The Institutional Review Board waived the requirement to obtain informed consent. No exclusion criteria were applied. Data collected included age, coexisting airway abnormalities, comorbid conditions, APGAR scores, sleep study results, parental and provider reported signs and symptoms, time to extubation (days), reoperation rates, length of follow up (calculated from date of surgery) and post-operative complications. If the child had ever been diagnosed with GERD by a physician prior to surgery then this was counted as the presence of this condition. Weight and height percentiles were calculated based on the growth chart for children with Down syndrome [8]. Sleep related symptoms included parental reports of noisy breathing, snoring or apneic pauses.

Procedures were performed by 3 staff surgeons. Patients were under general anesthesia, and the airway was visualized with the operating microscope. Cold laryngeal instrumentation or the microdebrider (or both) were used to divide the aryepiglottic folds and reduce redundant arytenoid tissue.

3. Results

3.1. Patient characteristics

Eighteen children were identified who had a diagnosis of Down syndrome and underwent supraglottoplasty for laryngomalacia (10 males, 8 females). Average age at surgery was 7.7 months (range, 0.6–25 months). Indications for surgery were feeding difficulties (n = 9 [50%]), noisy breathing or respiratory distress (or both) (n = 16 [88%]), sleep-related symptoms (n = 7 [39%]), and sleep apnea (AHI > 1) (n = 8 [44%]). The majority of patients (89%) had a coexisting airway lesion, most frequently subglottic stenosis (44%), pharyngomalacia (16%), tracheomalacia (44%), or bronchomalacia (22%). One patient had a paralyzed vocal cord. Gastroesophageal reflux was diagnosed in 11 patients (61%). Fourteen patients (78%) had congenital heart disease, including patent foramen ovale (33%), ventricular or atrial septal defect (44%), atrioventricular canal defect (11%), patent ductus arteriosus (17%), and coarctation of the aorta (11%).

3.2. Surgical outcomes

Sixteen patients (89%) were extubated by postoperative day (POD) 1 (6 on POD 0; 10 on POD 1). One patient remained intubated for 4 days because a cardiac procedure was performed concurrently with the supraglottoplasty. Description of post-operative symptoms was available for 16 patients. All of these parents reported decreased or resolved stridor. All but one parent reported improved feeding.

Weights were available for 16 out of the 18 children (89%). Nine children (50%) had an improvement in their weight percentile at a mean of 3.1 months (range, 1–6 months). However, half of patients did not demonstrate an improvement in their weight percentile after surgery. Average change in percentile for those who did improve was 17.6 percentile points and a loss of 16.7 points for those that did not improve. Overall change in weight percentile for all 19 patients was +2.6 points. Seven patients ultimately underwent adenotonsillectomy and 1 patient had adenoidectomy alone. Two patients later underwent tracheostomy and 2 required a gastrostomy tube. Two patients needed revision supraglottoplasty, and both had symptom improvement after the revision. Two patients died approximately 6 months after surgery. One child died at age 13 months (6 months after surgery) from ongoing difficulties with respiratory distress and recurrent respiratory infections due to severe laryngotracheobronchomalacia. The other patient died at age 9 months (6 months after surgery) at home of unknown causes (Table 1). We defined surgical failure of supraglottoplasty as lack of symptom improvement, the need for subsequent placement of a tracheostomy or gastrostomy tube, or the need for revision supraglottoplasty. By these criteria, the failure rate in our series was 33% (n = 6).

Preoperative and postoperative polysomnograms were available for 4 patients. All had a decrease in their postoperative apnea-hypopnea index (AHI) (median decrease = 6.5 points; range 5–58). Remarkably, one patient went from an AHI of 60 to 2 after supraglottoplasty alone. Postoperatively, all four of these patients were classified as having mild obstructive sleep apnea (OSA; AHI $\leq$ 5), although 1 patient had to undergo a revision supraglottoplasty to achieve this result. Three patients had postoperative complications. One child required nasal continuous positive airway pressure and high-flow oxygen during the first postoperative week and was eventually discharged on POD 12. One patient had increased work of breathing on POD 1 that responded to steroids and antibiotics. One patient was readmitted on POD 24 for aspiration pneumonia. The average duration of follow-up was 23 months (range 0–102 months).

4. Discussion

We present our experience treating children with Down syndrome (DS) and laryngomalacia during a 14-year period. Children with DS are more likely to have upper airway obstruction due to generalized hypotonia, narrow nasopharyngeal inlet, adenotonsillar hypertrophy, narrow midface and macroglossia [9]. As noted above, many of these children also have laryngomalacia. The etiology of laryngomalacia remains unknown although three main theories prevail. The anatomic and cartilaginous theories propose that the tissues around the supraglottis are redundant or flaccid

Table 1
Surgical outcomes.

	n	%
Total patients	18	100
Improved weight percentile	9	50
Subsequent adenotonsillectomy or adenoidectomy	8	44
Revision supraglottoplasty	2	11
Subsequent tracheostomy	2	11
Subsequent gastrostomy tube	2	11
Deaths (not surgery related)	2	11