RECURRENT CROUP AND PERSISTENT LARYNGOMALACIA: CLINICAL RESOLUTION AFTER SUPRAGLottoplasty

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PURPOSE: Recurrent croup (RC) is a common condition affecting the pediatric airway. This study seeks to determine if children with RC and persistent laryngomalacia (LM) clinically improve after supraglottoplasty (SGP).

METHODS: A retrospective review (2011-2014) of prospectively collected database of children with LM cared for at a single tertiary children’s hospital. Children with diagnosis of RC (>2 episodes of spasmodic croup in a season requiring steroids) and documented LM that underwent SGP were identified. Clinical history, demographics, clinical outcomes including episodes of croup before and after SGP, and operative complications were reviewed. Table 1

RESULTS: Out of 107 patients undergoing SGP for LM, 6 patients (5.6%) were diagnosed with RC. Mean age at first croup episode was 11.5 months. Mean age at SGP was 4.3 years. Figure 1. Mean number of emergency department visits was 3.2 (range 2-6 visits) prior to SGP. Mean number of episodes of croup requiring systemic steroids before and after SGP was 9.8 vs. 0.2 (p=.003). Mean length of followup after SGP was 24.5 months (range 12-40 months). There were no postoperative complications.

Conclusion: This is the first series to describe the clinical resolution of croup episodes in children with LM corrected by SGP. Recurrent croup should be added among other conditions associated with late-onset or persistent laryngomalacia.

DISCUSSION

Laryngomalacia (LM) is the most common laryngeal anomaly and source of stridor in newborns. LM describes the flaccid laryngeal tissue and inward collapse of supraglottic structures that occur during inspiration. In infants the severity of LM can be variable with retractions, intermittent cyanosis or apnea, cough, choking, feeding disruption, and reflux. Most cases of LM respond to treatment with expectant feeding modification and reflux therapy, with resolution by 12-24 months. However, up to 20% of infants with LM may progress to severe disease requiring surgical treatment. Supraglottoplasty (SGP) is a proven technique involving endoscopic trimming of the aryepiglottic folds and removal of redundant soft tissue overriding the arytenoids.1 SGP successfully resolves symptoms in 75-94% of patients.

A separate cohort of patients with flexible endoscopic findings of LM, but occurring at an older age has been coined as, late-onset laryngomalacia.2 In these patients, the symptom complex is often different from that of patients with congenital disease. Described as “state dependent” LM, patients will demonstrate obstruction at the arytenoid and supra-arytenoid levels during the disease “state”.3 Conditions reported include feeding-disordered LM, sleep-disordered LM, and exercise-induced LM.

This study describes another “state dependent” form of LM in the setting of recurrent croup (RC). Approximately 6.4% of children will experience multiple episodes of croup in the first 4 years of life. RC is not a specific diagnosis in itself, but rather should alert the clinician to identify a number of possible underlying causes. This workup commonly includes investigation for gastroesophageal reflux (GER), allergy, asthma, and congenital or acquired anatomic airway abnormalities. If there are clinical findings consistent for GER, anti-reflux therapy can be helpful. Hoa et al reported a significant decline in the number and duration of croup episodes in 87% of their patients.4 Given that up to 80% of infants with LM have GER, it would be reasonable to suspect clinical improvement in croup episodes in our patient population with GER therapy. However, 4 of the 6 current patients that did have clinical symptoms of GER and were treated with empiric anti-reflux therapy had no reduction in croup episodes despite appropriate duration of therapy. This would indicate that the LM was the underlying mechanism for the RC rather than untreated GER. Direct laryngoscopy and rigid bronchoscopy can be used to detect anatomic airway abnormalities.

In the current study the average number of episodes of croup requiring systemic steroids before and after SGP was 9.8 vs. 0.2 (p=.003). This indicates a clinically significant reduction with this intervention. Additionally, the average number of ED visits was 3.2 and average time from initial diagnosis to SGP was 3.4 years. All of these variables point to a burden of disease both in length and severity that was alleviated by SGP. This is the first clinical series of patients to describe recurrent croup as an entity of “state dependent” LM. One previous case report, highlights a patient with LM and RC with symptoms that resolved after SGP.5 In that report, the patient had clinical and endoscopic evidence of mild LM diagnosed at 3 months of age. The patient was observed and the stridor resolved. However, the patient began to develop RC after 1 year of age and underwent SGP at age 2. In our study, 2 patients had clinical evidence of stridor and LM as an infant that also resolved. Thus, this “state dependent” entity may be better described as persistent LM, rather than late-onset LM. The endoscopic findings of foreshortened aryepiglottic folds and prominent supra-arytenoid tissue at the time of SGP support this hypothesis. Figure 1

While clinically asymptomatic from the LM at baseline, we hypothesize that the underlying upper respiratory infection generates supraglottic inflammation which then exacerbates the LM. As a result, symptoms of stridor, barky cough, dyspnea manifest in RC secondary to prolapsing of the supra-arytenoid tissue. The evidence of clinical resolution of symptoms following SGP despite multiple previous interventions also supports this hypothesis. In conclusion, in a child with RC and clinical history or symptoms consistent with LM, flexible laryngoscopy can confirm the diagnosis. Patients with LM and RC croup that have failed previous medical interventions can significantly benefit from SGP with symptom resolution.