Laryngomalacia in Patients with Craniosynostosis

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Background

Craniosynostosis is described as the premature fusion of one or more cranial sutures and can result in a disfigured skull shape and increased intracranial pressure. Consequent neurodevelopmental aberrations[1] could increase these patients' risk for laryngomalacia as craniosynostosis has been described to occur with laryngomalacia in many cases with high rates of respiratory difficulty and airway abnormalities evident in patients with syndromic craniosynostosis.[2-4]

Laryngomalacia is characterized by altered laryngeal sensorimotor integration and tone resulting in dynamic supraglottic tissue prolapse,[5] typically presenting as stridor during the first few weeks of life with gradual recovery by 18-24 months of life with treatment of associated gastroesophageal reflux disease (GERD). Neurologic dysfunction has been observed in 20% of infants with laryngomalacia, with these patients presenting with greater severity of laryngomalacia.[5]

Objectives

- To characterize differences in the clinical presentation and treatment of laryngomalacia in patients with craniosynostosis compared to the general population
- To identify characteristics of patients with craniosynostosis that may increase the risk and severity of laryngomalacia.

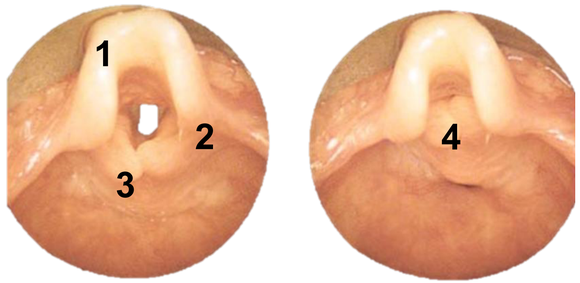


Figure 1: Epiglottis in a patient with laryngomalacia.
(1) omega shaped epiglottis
(2) shortened aryepiglottic folds
(3) redundant arytenoid mucosa
(4) inspiratory prolapse of arytenoid mucosa

Adapted from J Laryngol Otol 1977 Oct;91(10):887-92

Methods

- The control group was assembled from a database search performed on patients seen at Children's Hospital of Pittsburgh of UPMC (2007-2012) with isolated laryngomalacia.
- The experimental group was assembled from a database search performed on patients seen at Children's Hospital of Pittsburgh of UPMC (2000-2016) with concomitant laryngomalacia and craniosynostosis.
- Chart review and data collected included severity of laryngomalacia, comorbidities, symptom presentation and resolution, treatments and associated complications, polysomnography (PSG) results, and type of craniosynostosis (simple, multisutural, syndromic).
- Information regarding swallow function was obtained from clinical symptoms, clinical feeding evaluation studies (CFE), modified barium studies (MBS), and fiberoptic endoscopic evaluation of swallowing studies (FEES).

Conclusions

- Isolated laryngomalacia classically presents as inspiratory stridor; however, patients with craniosynostosis and concomitant laryngomalacia presented with stridor or stertor.
- Additional airway complications of patients with craniosynostosis presenting with laryngomalacia included tracheomalacia, bronchomalacia, subglottic stenosis (SGS), and obstructive sleep apnea (OSA) requiring additional interventions such as tracheostomy and CPAP.
- Swallowing dysfunctions, including dysphagia with aspiration and failure to thrive (FTT), are exacerbated in patients with craniosynostosis presenting with laryngomalacia, precipitating the need for gastrostomy and nasogastric (NG) tubes.
- Presentations, complications, and treatments of laryngomalacia are significantly different in the context of craniosynostosis.

Results

Table 1: Presenting Symptoms

	Isolated (n=68)	CS (n=32)	p-value
Stridor, % (n)	91.2 (62)	68.8 (22)	<0.01
Stertor, % (n)	11.8 (8)	34.4 (11)	0.01
Dysphagia, % (n)	17.7 (12)	50.0 (16)	0.002
Aspiration, % (n)	1.5 (1)	37.5 (12)	<0.0001
FTT, % (n)	7.4 (5)	34.3 (11)	0.001
OSA, % (n)	4.4 (3)	25.0 (8)	0.004

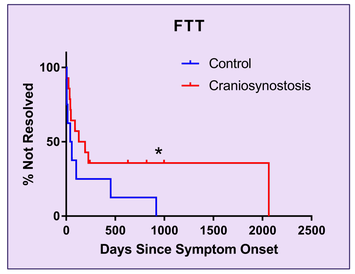
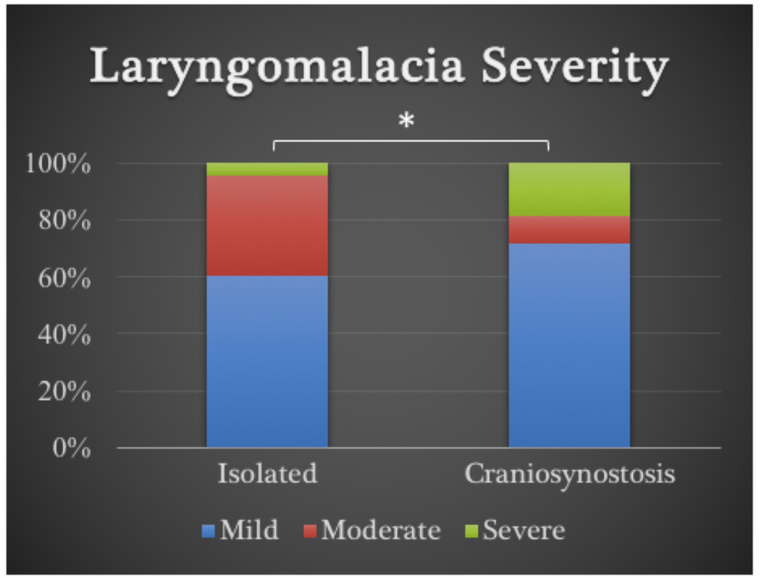


Table 2: Other Airway Abnormalities

	Isolated (n=68)	CS (n=32)	p-value
Tracheomalacia, % (n)	7.4 (5)	31.3 (10)	0.005
Bronchomalacia, % (n)	1.5 (1)	18.8 (6)	0.004
SGS, % (n)	22.1 (15)	43.8 (14)	0.03
Laryngeal Cleft, % (n)	1.5 (1)	0.001	0.001
BOT Collapse, % (n)	4.4 (3)	40.6 (13)	<0.001

Table 3: Treatments Utilized

	Isolated (n=68)	CS (n=32)	p-value
Gastrostomy Tube, % (n)	0.0 (0)	50.0 (16)	<0.001
NG Tube, % (n)	0.0 (0)	31.3 (10)	<0.001
Tracheostomy, % (n)	0.0 (0)	21.9 (7)	<0.001
CPAP, % (n)	0.0 (0)	31.3 (10)	<0.001



Future Directions

Pending data to be analyzed include the following:

- Effects of the following on symptom resolution:
  - GERD medications
  - Thickening of feeds
  - Supraglottoplasty
- Differences in presentations and outcomes between simple, multisutural, and syndromic craniosynostosis
- Differences in pre/post-tx CFE, MBS, and FEES
- Differences in pre/post-tx PSG results

A follow-up study will investigate the prevalence of total airway anomalies in patients with craniosynostosis and attempt to identify characteristics of craniosynostosis that may increase the risk of development of airway anomalies.

References

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Pennsylvania Academy of Otolaryngology 2017