Multidisciplinary Management of Congenital Subglottic Stenosis in the Setting of Cystic Fibrosis

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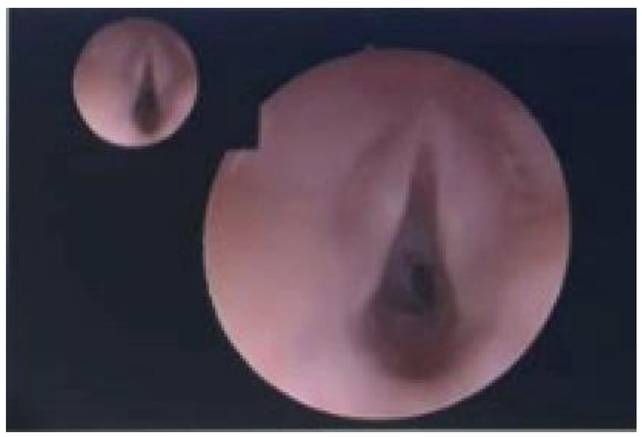
BACKGROUND

Subglottic stenosis (SGS) is a common laryngeal anomaly in infants and can be congenital, acquired, or acquired upon congenital. To date there have been no case reports of congenital SGS in the setting of cystic fibrosis (CF). This case report highlights the unified airway theory of CF and the need for multidisciplinary management of a congenital airway anomaly in this context.

CASE PRESENTATION

4-month-old male with homozygous delF508 CF who has required multiple hospitalizations for CF exacerbations complicated by multiple transfers to the PICU and failed attempts at extubation.



First MLB showing elliptical cricoid with grade 3 SGS

INTERVENTIONS AND TIMELINE

DIAGNOSTICS

1) ENT consult → concern for cord injury from intubations, mild arytenoid edema on flexible scope

2) ENT consult → new stridor, diagnosed mild LM on flexible scope

3) ENT consult → worsening biphasic stridor, MLB showed elliptical cricoid with grade 3 SGS

MULTIDISCIPLINARY MANAGEMENT

ENT: MLB and balloon dilation every 2 weeks to manage airway edema from acute infections

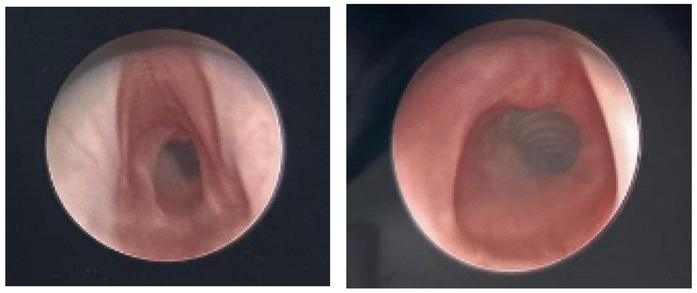
Pulmonology: Cystic fibrosis and antibiotic management

Nutrition: Diet and pancreatic enzyme replacement

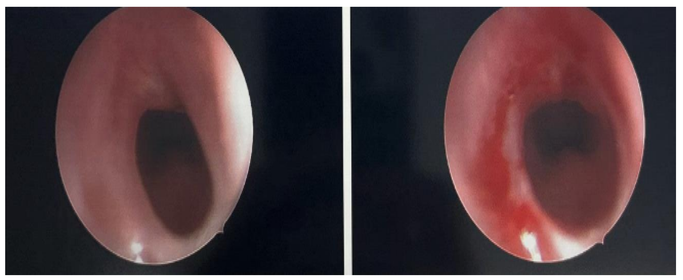
LARYNGOTRACHEOPLASTY

Single stage LTP using anterior thyroid ala cartilage graft

1-week post-operative MLB showed well healing graft with thin scar band dilated with endotracheal tube



One-week post-operative MLB following laryngotracheoplasty



Three-month surveillance MLB following laryngotracheoplasty

CONCLUSIONS

This case emphasizes the unified airway theory and need for a broad differential of airway diagnoses in patients with CF who require repeated hospital admissions, unexplained O2 requirement, and intubation. A multidisciplinary approach is critical in the management of these complex airway patients.