Tracheal Stenosis in Morquio A Patients: Identification, Treatment and Perioperative Events
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Introduction
• Tracheal stenosis is a novel finding and previously unrecognized (1) in patients with Morquio syndrome.
• This study was conducted to assess severity of tracheal stenosis in our cohort of Morquio A patients who had respiratory symptoms or tracheal narrowing seen in MRI studies of cervical spine(2). Further, study describes the surgical treatment and peri-operative events patients underwent.

Methods
• IRB approved retrospective study
• Subjects had progressive obstructive symptoms or evidence of airway obstruction in their cervical spine MRI(2).
• Subjected to CT Angiogram and pulmonary evaluation to evaluate large airway obstruction.

Results
• 44 patients included in study
• Average age, weight and height (12.3yrs, 102.5cm, 23.5kg)
• 7 of 44 have completed evaluation of their tracheal stenosis (Figure 1)
• Tracheal stenosis was severe in all but one patient.
• Three patients had surgical correction with tracheal resection by cardiothoracic surgery (Table 1)
• 6 of 7 patients were known difficult airway (both mask ventilation and intubation)
• 6 of 7 patients intubated with 5.5cm cuffed ETT or smaller
• Only one patient showed obstructive pattern on PFT while the rest showed restrictive lung disease.

Discussion
• Life threatening tracheal stenosis should be suspected and evaluated for when increasing respiratory symptoms are present.
• Symptoms are often mistaken for upper airway symptoms caused by glycosaminoglycan (GAG) deposits.
• Surgical intervention may be the definitive treatment to relieve tracheal obstruction.
• Physicians must take into account the severity of respiratory symptoms and underlying pathology.
• Multidisciplinary team approach should be used when caring for these complex patients.

Conclusions
• Many anesthesiologists perform anesthetics without realizing the risk and underlying respiratory issues in these patients.
• Our study emphasizes need for recognition and evaluation of complex airway pathology prior to all anesthetics.
• Perioperative safe conduction of anesthesia may emerge as an indication for intervention to relieve tracheal stenosis preemptively prior to anesthesia in Morquio patients.

References
1. Theroux et al: Pediatric Anesthesia 2012
2. Tomatsu: Molec genetics and metabolism 2015.

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