WHEN LARYNGOMALACIA ISN’T LARYNGOMALACIA

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Introduction:
Compression of the pediatric airway can be an often unrecognized complication of congenital cardiac and aortic arch anomalies. A high index of suspicion should be maintained in infants and children with recurrent respiratory difficulties, stridor, wheezing, dysphagia, or apnea unexplained by other causes. Vascular rings are formed by abnormal persistence and/or regression of components of the aortic arch complex. These structures can cause compression of the trachea, bronchi, and esophagus. Although accounting for less than 1% of congenital heart defects, vascular rings are important sources of airway and esophageal obstruction.

Case #1:
A full term infant boy with a past medical history significant for microcephaly and a chromosomal translocation initially presented to ENT clinic with noisy breathing. The patient was diagnosed with mild laryngomalacia. He was given a short course of steroids, which did not improve his symptoms. At the patient’s 6 month follow-up visit he was noted to have worsening stridor, especially with feeding. The patient subsequently underwent a rigid bronchoscopy with ENT and was noted to have tracheal compression of the right anterior wall 2 centimeters proximal to the carina.

The patient underwent CT angiography, which demonstrated evidence of a very subtle compression of the trachea. The patient was scheduled for cardiac surgery and in fact found to have an aberrant brachiocephalic trunk causing tracheal compression. The patient underwent anterior re-implantation of the brachiocephalic trunk in the ascending aorta.

The patient subsequently received a CT angiography as well as a bronchoscopy on the day following admission revealed distal tracheal compression by an anterolateral mass 3 cm proximal to the carina. There was no significant laryngomalacia noted and no foreign body recovered. The patient subsequently received a CT angiography as well as a cardiac catheterization which revealed a vascular ring surrounding the trachea and esophagus. The patient was taken to the operating room by cardiac surgery for repair.

Case #2:
A 15 month old girl with a past medical history significant for laryngomalacia, complicated by coughing and gagging with every meal, presented to the ED after an episode of apnea, unresponsiveness, and cyanosis after choking on a sausage at breakfast. She was resuscitated in the field and admitted to the hospital for observation and workup. Of note, the patient had been diagnosed with laryngomalacia at 2 months of age. She had been prescribed albuterol 3x/day without improvement of her symptoms. She also had multiple ED visits for recurrent URI symptoms.

A bronchoscopy by ENT on the day following admission revealed distal tracheal compression by an anterolateral mass 3 cm proximal to the carina. There was no significant laryngomalacia noted and no foreign body recovered. The patient was taken to the operating room by cardiac surgery for repair.

Discussion:
The goal of operative repair is to divide the compressive vascular ring, relieve tracheal and/or esophageal compression, and maintain normal perfusion to the aortic arch. Cartilage destruction and malacia of the infant airway have been observed to appear within weeks of induced external compression. Early repair may reduce airway morbidity and allow for normal tracheobronchial growth.

In terms of anesthetic management, it may be advisable to approach these patients with the same considerations as patients with anterior mediastinal masses. Spontaneous ventilation and avoidance of muscle relaxation may be beneficial in terms of maintaining airway patency during inhalational induction. In most cases, the endotracheal tube should be placed so that it lies above the stenotic segment to avoid edema in the narrowed area.

Successful tracheal extubation is possible in most patients with isolated vascular rings at the conclusion of the procedure. It is important to remember that successful repair of the ring may not immediately relieve airway obstruction. Residual obstruction may be observed immediately on extubation. Bronchoscopy performed at the end of surgery may be helpful to assess the effectiveness of surgery. Recurrent laryngeal nerve injury can occur and carries the risks of stridor, hoarseness, and potential for aspiration.

References: