AB014. Tracheomalacia

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Abstract: Tracheomalacia is a rare condition that is poorly understood. It typically is characterized by weakness of the tracheal cartilages and reduction and atrophy of the elastic fibers of the membranous wall. It typically involves the intrathoracic trachea and oftentimes both main stem bronchi. This is not to be confused with conditions such as Mounier-Kuhn syndrome or polychondritis. It is also not to be confused with post-intubation or post-tracheostomy malacia, COPD, relapsing polychondritis or extrinsic compression. The incidence of tracheomalacia is unknown but thought to be quite uncommon. It is less than 1% of 1,500 patients who underwent bronchoscopy for a variety of reasons and this figure may be as high as 5% as reported by others. It is a condition that predominantly involves men and generally in an age group older than 50. The symptoms of tracheomalacia are dyspnea, barking cough (barking seal cough), difficulty clearing secretions with resultant recurrent infections, choking or syncopal spells related to coughing, difficulty in extubation following surgery and is typically worse on emergence from anesthesia. The diagnosis of tracheomalacia is challenging. Oftentimes the chest X-ray is normal. Bronchoscopy is the gold standard in making this diagnosis and should show a reduction in the AP diameter of the trachea by 50% or greater. Awake bronchoscopy with provocative maneuvers such as deep breathing and coughing should demonstrate this finding as well as forward displacement of the membranous wall with an archer's bow deformity of the cartilage. A provocative maneuver of inserting a Y stent has been advocated by some to determine whether or not the condition is true tracheomalacia versus COPD. In true tracheomalacia the insertion of a Y stent should lead to fairly immediate improvement in the ventilation. If such a finding is demonstrated, this is confirmatory evidence that the underlying problem is predominantly tracheomalacia. It does not exclude the coexistent presence of COPD. Pathology in patients with tracheomalacia usually shows the gross findings of malacia with AP narrowing, a widening and flaccid membranous wall with atrophy of the longitudinal elastic fibers. Sometimes there can be evidence of fragmentation of the tracheal cartilage. Radiologic evaluation: Dynamic CT scan with inspiratory and expiratory phases is quite helpful. On standard inspiratory CT scans it may be normal, but forced exhalation CT oftentimes reveals reduction in the diameter of 80% or more and is only 35% in normal patients. Finding of greater than 50% narrowing on dynamic CT is thought to be diagnostic. The addition of virtual bronchoscopy may add to the evaluation of patients with tracheomalacia. Pulmonary function testing in tracheomalacia is helpful but not diagnostic. Reduced FEV1 and PEFR with a rapid decrease in flow and preservation of the shape of the inspiratory limb is characteristic. The expiratory limb of a flow volume loop is very flat. Unfortunately the findings in tracheomalacia are very similar to COPD and therefore not pathognomonic of this underlying condition. Treatment of tracheomalacia: if the condition is mild and the symptoms are mild, no treatment is required. Supportive care of underlying COPD should be aggressively instituted. CPAP may improve FVC and improve dynamic collapse and might be of value in managing patients without surgery. T-tubes with extra-long distal limbs have been advocated by some if the tracheomalacia is limited to the trachea. This obviously involves an appliance and the sidearm of the T-tube exiting the neck. Patients generally wish to avoid that. Internal stents have been tried, but oftentimes associated with coughing, migration, secretions, tracheitis, bronchitis and pneumonias, and therefore have a very limited role. Herzog and Nissen advocated plication of the membranous wall and some type of rigid material to pull the two ends of the splayed tracheal cartilages together to achieve a more normal configuration. This concept has been further refined by others, but the basic principles remain. When surgery is done, the basic concepts put forward by the Nissen and Herzog have remained. Most commonly is some type of mesh cut to the size of the desired diameter of the membranous wall and secured to the ends of the tracheal cartilage. In addition, the membranous wall is sutured to this mesh material to provide stabilization of the membranous wall and eventually integration of the membranous wall and the mesh. Once these things have been done, the mesh is secured in place thereby reducing the overall diameter, reconfiguring the trachea in the normal anatomic shape and fixation of the membranous wall. If the process involves the main stem bronchi, they also need to undergo the same type of procedure. Small series at the Massachusetts General Hospital employing the previously described technique has led to satisfactory results. Patients were diagnosed by bronchoscopy and CT scan. Symptoms were typical of patients with tracheomalacia and the indications

for surgery were for intractable symptoms unresponsive to medical treatment including stent placement. Fourteen patients were in this early series, mean age of 54, 11 of 14 were men, eight had underlying COPD. The length of stay in the hospital was eight days and three patients required postoperative ventilation between two and three days. One patient suffered pneumonia and another had a prolonged ileus. There were no deaths in the series. FEV1 improved from 51% to 74% and the PEFR improved from 49% to 70%. There was corresponding improvement in flow volume loops following tracheoplasty. Long-term results in this group of 14 patients showed that there were two late deaths, one from COPD and the other from an unrelated cause. One patient who had a good result for five years then had progressive COPD with an intact repair. Six had excellent results, two were good and two were failures with progression of their bronchomalacia. The Beth Israel Deaconess Hospital in Boston has also had an experience treating this condition. They have employed the use of Y stents as a diagnostic maneuver more than others and recommend this in the evaluation and treatment of tracheomalacia. Their series are very similar to the MGH series with similar results confirming the value in patients with diagnosed tracheomalacia. They also emphasize the difficulties in distinguishing COPD from tracheomalacia and in those cases have found the use of these Y stents to be very helpful. In summary, tracheomalacia is underdiagnosed. The symptoms are very typical with cough, dyspnea, inability to clear secretions and recurrent infections. It is very difficult to distinguish COPD from tracheomalacia and is the basis of the issues regarding patient selection. Dynamic CT scans and bronchoscopy are essential in the diagnosis and patient selection. Y stents can be helpful in determining those who will have a favorable response to posterior tracheal splinting. The initial treatment is supportive, but when confident of the diagnosis posterior wall tracheoplasty provides effective symptomatic relief.

Keywords: Tracheomalacia; posterior tracheobronchoplasty

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