Laryngomalacia, Tracheomalacia and Bronchomalacia

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Airway malacia can occur in the larynx (larygomalacia), trachea (tracheomalacia), or bronchi (bronchomalacia). As a group these are the most common congenital abnormalities of the pediatric airway and are characterized by increased airway compliance, resulting in excessive dynamic collapse during the respiratory cycle. While a diagnosis can be suspected based on clinical history and physical examination, definitive evaluation is based of nasopharyngolaryngoscopy and/or bronchoscopy. Observation and conservative management are typically all that are required. However, surgical intervention can be necessary in the most severe cases, and can result in significant improvement in symptoms.

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Dynamic Airway Collapse

Malacia refers to an excessive softness of a tissue, most often a bone or cartilage. In the case of the airway, malacia refers to increased compliance of the conducting airways. Airway malacia can occur at the level of the larynx (laryngomalacia), trachea (tracheomalacia), bronchi (bronchomalacia), or permutations of all three.

Malacia is the most common congenital abnormality of the pediatric airway.

Laryngomalacia

Epidemiology

Laryngomalacia is the most common congenital anomaly of the larynx and accounts for more than 60% of the cases of stridor in pediatric patients.[1,2] Supraglottic structures collapse into the larynx during inspiration and result in impendence to airflow. The larynx may be obstructed by prolapse of the aryepiglottic folds medially; the arytenoid, corniculate, or cuneiform cartilage anteriorly; the epiglottis posteriorly; or a combination of the three (Fig. 1A and D).[3]

Intervention is only required in the most severe cases.

The precise etiology of laryngomalacia is not well understood. Historically, abnormalities of the laryngeal structures and cartilage have been postulated to cause laryngomalacia. Indeed, some patients with severe disease have a shortened aryepiglottic fold to glottic length ratio; however, similar decreased ratios can be seen in patients without laryngomalacia. Histologic specimens of laryngeal cartilage demonstrate mucosal edema and dilation of the lymphatic vessels, but no chondropathy.[4] More recent research has identified abnormalities in sensorimotor integration in children with laryngomalacia.[5,6]

While laryngomalacia has been associated with neurologic, genetic, and cardiac disorders, gastroesophageal reflux disease is the most common comorbidity.[7] Theoretically, breathing against and obstructed airway causes worsening reflux and laryngeal edema, which exacerbates airway obstruction, creating a cyclical process and worsening clinical symptoms.

Signs and Symptoms

Positional stridor is present in the majority of infants with laryngomalacia, though it may not be the primary reason for presentation.[8,9] Other respiratory symptoms such as accessory muscle use, respiratory distress, and hypoxemia can also occur, but are more common in severe cases. Patients with severe disease are also at increased risk for obstructive sleep apnea. Patients may also develop non-respiratory symptoms, especially in

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severe disease. Feeding difficulty and dysphagia are frequently seen, and the combination of inadequate intake from impaired feeding and increased metabolic demand from respiratory distress can lead to failure to thrive.[8,9]

Symptoms usually begin within the first several days of life, but children may not present to medical attention until several months of age.[8] Respiratory symptoms are typically worse in the supine position or during periods of increased respiratory effort such as crying or feeding. While symptoms can get worse over the first several months of life, patients typically have resolution by two years of age and often earlier.[2,8]

Diagnosis

Laryngomalacia can be suspected based on the classic findings of positional inspiratory stridor. However, other pathology can be indistinguishable from laryngomalacia based solely on clinical history and physical examination,[10] and radiographic techniques demonstrate poor accuracy in identifying disease.[11] Thus, direct visualization with nasopharyngolaryngoscopy (NPL) is the gold standard for the diagnosis of laryngomalacia. NPL can be performed in an awake patient seated upright in a parent's arms[12]; however, sedated NPL in the supine position with spontaneous respiration is more sensitive and specific for establishing the diagnosis.[13]

Definitive diagnosis require direct airway visualization.

Treatment

Laryngomalacia typically has a benign course and resolves without interventions. Observation and/or conservative treatments with positioning and feeding therapy are sufficient in ~90% of patients.[8] In patients with severe respiratory symptoms or failure to thrive, surgical intervention may be necessary.[14] Supraglottoplasty is the most commonly performed surgical intervention and is successful for symptomatic relief and improved weight gain in most patients.[15] In patients with posterior collapse of the epiglottis, epiglottopexy can be used to ameliorate symptoms.[16] If endoscopic surgery is unsuccessful, tracheostomy is an alternative treatment for refractory disease.[17]

Tracheomalacia

Epidemiology

Tracheomalacia is characterized by an increased compliance of the intra or extrathoracic trachea resulting in dynamic collapse during the respiratory cycle (Fig. 1B and E). Congenital tracheomalacia results

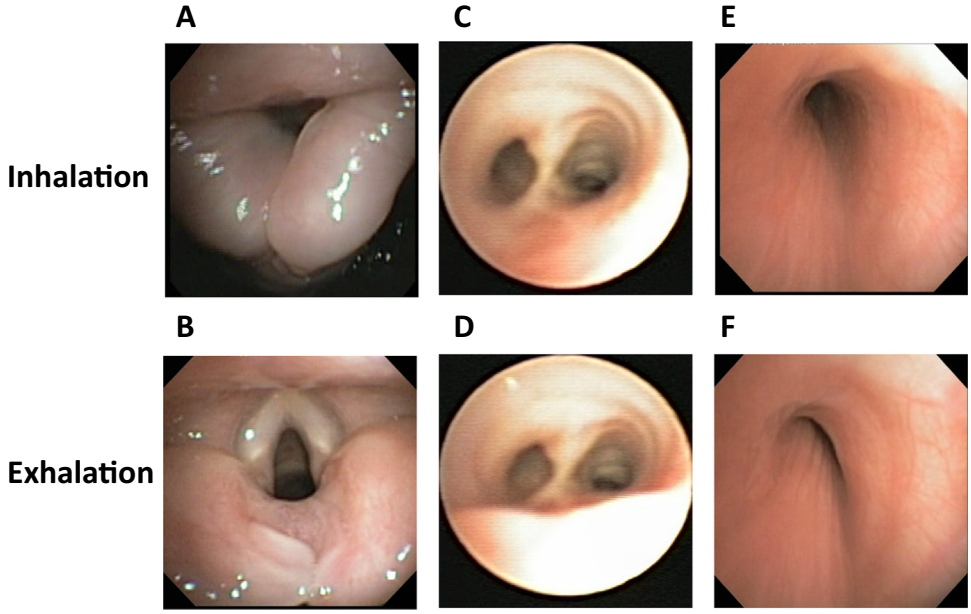

Figure 1. Endoscopic view of the larynx during inhalation with anterior prolapse of the arytenoid cartilage in a patient with laryngomalacia (A) and during exhalation (B). Endoscopic view of the distal trachea during inhalation(C) and during exhalation with partial collapse of the tracheal lumen in a patient with tracheomalacia (D). Endoscopic view of the left main bronchus during inhalation (E) and during exhalation (F) with nearly complete occlusion of the lumen in a patient with bronchomalacia.