Tracheobronchomalacia in Adults

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Severe, diffuse tracheobronchomalacia (TBM) is an underrecognized cause of dyspnea, recurrent respiratory infections, cough, secretion retention, and even respiratory insufficiency. Patients often have comorbidities, such as asthma or chronic obstructive pulmonary disease, and inappropriate treatment for these conditions may precede eventual recognition of TBM by months or years. Most of these patients have an acquired form of TBM in which the etiology in unknown. Diagnosis of TBM is made by airway computed tomography scan and flexible bronchoscopy with forced expiration. The prevailing definition of TBM as a 50% reduction in cross-sectional area is nonspecific, with a high proportion of healthy volunteers meeting this threshold. The clinically significant threshold is complete or near-complete collapse of the airway. Airway stenting may treat TBM, although complications resulting from indwelling prostheses often limit the durability of stents. Surgical stabilization of the airway by posterior splinting (tracheobronchoplasty) effectively and permanently corrects malacic airways. Proper surgical selection is facilitated by a short-term stent trial.

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Tracheobronchomalacia (TBM) is a condition characterized by excessive weakening of the walls of the trachea and bronchi. Malacic tracheobronchial airways demonstrate dynamic collapse with expiratory maneuvers.[1] Although airway malacia may present as isolated, segmental weakness of the trachea, or less commonly of the bronchi alone, the scope of this article is the severe, diffuse form of acquired TBM that affects the central airways. This manifestation of TBM is an acquired condition, and progression to its diffuse form is suggested by a study of bronchoscopic surveillance of patients with either isolated tracheomalacia or bronchomalacia in whom 67% and 100% of the patients, respectively, evolved to diffuse TBM during a mean follow-up interval of 5 years.[2]

There is increasing recognition of TBM in patients with respiratory complaints, although the true incidence of TBM in the adult population as a whole remains poorly understood. TBM previously has been reported in 4.5% of the general population.[3] Ikeda et al[4] showed that more than 13% patients who were undergoing evaluation for respiratory complaints were found to have TBM. Estimates as high as 23% have been reported in patients with a diagnosis of chronic bronchitis.[5] In the pediatric population, the incidence of primary congenital TBM is difficult to assess because evaluation of infants with respiratory complaints may yield high rates of detection of TBM.[6] After reviewing their institution's series of pediatric bronchoscopies, Boogaard et al[7] estimated the incidence of airway malacia at 1 in 2100 live births.

A main reason why the incidence of TBM in the general population remains ill-defined is that the extent of airway collapsibility required to meet the threshold for pathologic collapse is not well understood. The standard definition is greater than 50% reduction in airway cross-sectional area with expiration. However, when patients with emphysema were examined, for example, more than 13% were found to have TBM by the standard definition, but when the threshold was raised to greater than 70% airway collapse, only 5% of this cohort met the definition of TBM.[8] Recent work from our institution has demonstrated that 78% of healthy, asymptomatic volunteers may exceed the threshold of collapsibility for the diagnosis of TBM, with a mean reduction in tracheal expiratory cross-sectional area of nearly 55% in these study subjects.[9] The implication is that a normal tracheobronchial tree will exhibit physiological degrees of collapse. Thus, when evaluating patients with TBM at Beth Israel Deaconess Medical

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Center (BIDMC), we restrict the nomenclature of severe TBM to patients with complete or near-complete collapse (>90%) of the airways.

CLASSIFICATION

Broadly, TBM can be classified as congenital or acquired, in addition to designations of diffuse or localized disease. In the pediatric population, tracheomalacia may be associated with tracheoesophageal fistula, and there is some controversy whether this localized malacia represents acquired or congenital disease.[10-13] Other conditions associated with TBM are matrix disorders that diffusely impair collagen formation and tracheal wall development, including polychondritis, chondromalacia, and mucopolysaccharidoses.[6,14-17] Other abnormalities of matrix biology and even genetic predispositions also deserve further examination.[18,19] Acquired TBM in the pediatric population may result from a host of pathologies that may also be seen in the adult population, including vascular anomalies, goiters, and tracheotomy, all of which result in localized forms of malacia.[11] The diagnosis and treatment of TBM in the pediatric population is outside the scope of this article but has been the subject of an excellent review by Carden et al.[11]

Most adult patients diagnosed with TBM have acquired forms, although a smaller subset of patients exists who have tracheobronchomegaly (Mounier-Kuhn syndrome) but do not present until adulthood.[20-22] Tracheobronchomegaly is diagnosed when the transverse diameters of the trachea, right main stem bronchus, and left main stem bronchus exceed 3.0, 2.4, and 2.3 cm, respectively, and obviously represents a diffuse form of malacia.[23] The putative causes of acquired forms of TBM may be divided into inflammatory and compressive etiologies, such as tracheotomy and endotracheal intubation, trauma and lung surgery (eg, pneumonectomy and post-pneumonectomy syndrome), relapsing polychondritis (RP), vascular abnormalities, goiters, and even exposure to toxins, such as mustard gas.[24-26] In most adult patients evaluated at our institution, however, the actual cause of their TBM is not known.

Morphologic classification of TBM has been described by several investigators.[26-28] Murgu and Colt[26] have proposed separating malacia that affects the cartilaginous wall of the airways into 3 categories: saber sheath-type TBM, circumferential-type TBM, and crescent-type TBM. They further characterize airways that only demonstrate membranous wall collapse as demonstrating either physiological collapse (dynamic airway collapse) or pathologic collapse (excessive dynamic airway collapse), but they differentiate this from true malacia. At BIDMC, we consider both cartilaginous and membranous structural collapse as forms of malacia because the more pertinent consideration for eventual success of a therapeutic intervention appears to be less whether it is the cartilaginous or membranous wall pathology that potentiates collapse but whether the presenting morphology is frown-shaped on expiration or lunate-shaped on inspiration, both of which are amenable to surgical stabilization.[27,29] Other types of TBM, such as primarily anterior or lateral cartilaginous found in idiopathic saber sheath tracheas or post-tracheostomy or concentric forms like RP, might not be adequately stabilized by tracheobronchoplasty.[24,26,30] Thus, given that either membranous or cartilaginous pathology may lead to symptoms and either may be treated by similar interventions, we do not see the need to separate the pure membranous intrusion into a separate nonmalacia diagnostic category of excessive dynamic airway collapse.

PRESENTATION

Adult patients with severe, diffuse TBM may present with dyspnea, recurrent infections, intractable cough, and retained secretions.[2,11,31] In addition, respiratory failure or failure of ventilator weaning may be the herald event that leads to a diagnosis of TBM.[31-33] More than 90% of patients who eventually move on to treatment for their TBM at our institution are found to have dyspnea as their significant complaint, either as a sole symptom or in combination with other symptoms.[33] This dyspnea is typically exertional, and in patients with severe collapse, the level of exertion may be minimal before the onset of symptoms. Often patients will complain of inability to climb a flight of stairs or even walk short distances on flat ground. In patients that we evaluate for consideration of intervention for their TBM, it is not uncommon to find that their activities of daily living, such as standing to shower or walking to a mailbox, are impossible to conduct. Patients with dyspnea may complain of wheezing, stridor, or noisy breathing, a sensation that their airflow is restricted on expiration, or an inability to inspire deeply. Some patients also will describe worsened dyspnea with bending or recumbency.

The cough associated with severe, diffuse TBM has a barking quality and may be paroxysmal in nature, triggered by minimal stimuli, such as laughing, exertion, or even speaking. Some patients will report that lying supine may also initiate coughing. The barking quality of the cough is presumably related to a reed-like, vibratory effect of the narrowed airway lumen. Syncope because of paroxysmal and unremitting coughing has been described.[34] In a study of nonsmoking patients with normal chest radiographs