REVIEW ARTICLE

TRACHEOBRONCHOMALACIA AND HYPERDYNAMIC AIRWAY COLLAPSE

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ABSTRACT

Tracheobronchomalacia (TBM) and hyperdynamic airway collapse (HDAC) are similar in presentation but differ in anatomy affected. These two entities refer to the loss of structural integrity of the airway walls. The prevalence of TBM and HDAC is not known. Diagnoses associated with TBM and HDAC include gastroesophageal reflux disease, chronic infections, emphysema, prior surgical manipulation of the trachea, sleep apnea, and collagen vascular diseases. Diagnostic testing includes computed tomographic (CT) scan of the chest with dynamic expiratory imaging, pulmonary function testing, six minute walk test, impedance probe for reflux disease, and a bronchoscopy for airway exam. If sleep apnea is suspected a polysomnogram should be pursued. If the inciting disease process can be identified, it should be aggressively treated. Weight loss is a critical aspect of the management of TBM and HDAC. Placement of a silicone stent into the airway may solidify the diagnosis and determine which patients will be successful with trachebronchoplasty.

INTRODUCTION

TBM and HDAC can be debilitating diseases which leave patients with significantly decreased functional capacities and an overall poor quality of life. The diverse etiologies behind TBM and HDAC make it difficult to diagnose and can delay an accurate diagnosis for years[1]. A thorough history of the illness is key to understanding the possible etiologies behind the disease as well as to guiding the diagnostic work up and to suggesting what treatments maybe most beneficial. This review will exclude the discussion of pediatric TBM as it is fundamentally different and is largely comprised of congenital diseases that rarely affect adults. Literature on TBM and HDAC are somewhat fraught with error[2]. Although there are considerable differences between TBM and HDAC, the strict definitions are not always used in the literature[2].

Description

TBM and HDAC are virtually indistinguishable by history and overlap considerably although they are different entities that often coexist. TBM refers to the loss of structural integrity of the cartilaginous structures of the airway wall[2]. The “soft” airway walls are mobile and lead to flattening of the normally curved cartilaginous structures with pressure changes that occur with coughing or forced expiratory maneuvers[1-2]. HDAC (also known in the literature as excessive dynamic airway collapse) is an exaggeration of the normal airway wall movement[2]. In normal individuals who have a structurally normal airway, the lumen of the airway can narrow up to 35% with coughing or forced expiratory maneuvers[2-3]. When the airway lumen decreases by more than 50% with forced expiratory maneuvers it is considered abnormal[1-2]. In HDAC airway compromise is due to invagination of the posterior membrane into the lumen of the airway[2]. In some cases, patients may have severe disease and have elements of incompetent cartilaginous structures as well as invagination of the posterior membrane on forced exhalation[1-2].

Prevalence

The prevalence of TBM and HDAC in the general population is not known. The prevalence in study populations vary dependent on the definition used as well as the type of test used to diagnose TBM or HDAC. In patients who are undergoing videobronchoscopy for evaluation of various respiratory complaints the incidence of TBM or HDAC ranges between 4 and 23%[1-2, 4-5]. Hasegawa and colleagues performed a retrospective evaluation of 163 consecutive patients who were being evaluated for suspected pulmonary embolism and found that 16 (10%) fit the criteria for TBM[6].

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Pathophysiology

The specific pathologic process is dependent on the etiology of TBM of HDAC. Literature regarding in depth insight into the histopathology of TBM is scarce although a few well described case series provides some insight[2]. In TBM the cartilaginous rings are the main sites of pathologic abnormality. In most cases the ratio of cartilage to soft tissue is decreased and in some cases absent[2]. The decrease or loss of cartilaginous tissue results in abnormal structural integrity. This loss of structural integrity is the key mechanism in TBM.

In HDAC the posterior membranous portion of the airway is redundant or lacks the ability to maintain normal tone[2]. Normally with inhalation the posterior membranous portion of the airway moves posterior and increases the luminal area of the airway. Longitudinal smooth muscle fibers tighten to prevent over distention. Upon exhalation the posterior membrane moves forward into the lumen of the airway. Longitudinal smooth muscle fibers again tighten to prevent excessive airway compromise. Available histopathology studies demonstrate that the smooth muscle fibers in patients with HDAC are decreased, thus allowing for exaggerated airway movements[1-2].

Patients with TBM and HDAC have dyspnea (accelerated with activity), cough, wheezing, and inability to effectively clear secretions, which predisposes to recurrent infections[1-2]. The ineffective mucociliary clearance as well as the predisposition to develop recurrent infections may create significant airway inflammation and perpetuate further damage to the airway.

Etiology

TBM and HDAC can occur in infancy and early childhood. The etiologies that are responsible for TBM and HDAC in infancy and early childhood differ from those that are associated with adult TBM and HDAC. Given the differences in etiologies, we will not discuss pediatric TBM and HDAC in this review. TBM and HDAC that develop in adults are acquired and often attributable to a known entity.

Several etiologies that cause TBM and HDAC are a result of airway inflammation. Prolonged intubation or the presence of a tracheostomy tube may result in weakening of the airway wall at the site of balloon inflation[7]. Inhalation or aspiration of chemicals or substances that incite irritation of the airway can weaken the airway wall. Examples of inhaled substances include tobacco smoke or the combustion of fossil fuels[2]. Examples of aspirated materials include the aspiration of gastric acid or aspiration of food. Inflammation of the airway soft tissue and cartilage can result from collagen vascular diseases including relapsing polychondritis[1-2, 8]. Chronic airway infections can be associated with intense airway inflammation and can lead to decreased airway wall integrity[1-2, 5]. Chronic obstructive pulmonary disease and asthma have been implicated in TBM and HDAC. The relationship with these common clinical entities is believed to be due to chronic airway wall inflammation as well as pressure changes in the chest promoting large airway collapse[1-2].

Mechanical injury or manipulation of the airway can be associated with TBM[1-2]. Closed chest trauma which results in fracture of tracheal cartilages can result in weakened airway walls. Airway or lung malignancies can directly destroy the airway wall or can mechanically compress the airway. Non-malignant chest tumors or masses (Thyroid goiter, aortic or pulmonary artery aneurysm) can also compress the airway leading to TBM. Patients who have had airway surgeries or endobronchial electrosurgery or laser therapy can develop focal TBM or HDAC.

Rare congenital diseases that are associated with TBM and HDAC can present in adulthood. Mounier-Kuhn syndrome (also known as congenital tracheobronchomegaly) is characterized by atrophy or absence of elastic fibers of the posterior membrane leading to large airways with minimal integrity[1]. Ehlers-Danlos syndrome is an inherited collagen vascular disease that is associated with weakened or lax cartilaginous structures leading to TBM[2].

Signs and Symptoms of disease

The signs and symptoms of TBM and HDAC are non-specific. Careful consideration and further testing should be sought for patients who do not respond as expected to usual and customary treatments for their initial diagnosis. Patients with TBM and HDAC may have an intractable cough, wheezing, dyspnea, and recurrent bronchitis or pneumonia[1-2]. Dyspnea and a cough that worsen with exercise are the principle complaints of patients who present with TBM and HDAC. Inhalation is generally not difficult for patients with TBM and HDAC whereas full exhalation is difficult and patients commonly complain that they cannot fully exhale.

Diagnosis

The diagnosis of TBM and HDAC is often delayed due to the non-specific symptoms and the overlap with many other respiratory illnesses[1]. Once suspected the approach to patients should be relatively uniform and directed based on the patient's history and findings from preliminary studies. Initial studies should include pulmonary function testing and computed tomography (CT) of the chest with dynamic expiratory imaging[1-2]. Pulmonary function testing serves to describe the physiological limitation of the airway and to exclude severe asthma or chronic obstructive pulmonary disease (COPD)[2]. Although asthma and COPD can occur with or be the cause of TBM and HDAC, judicial clinical rational should be exercised in the setting of mild TBM or HDAC with severe asthma or COPD. Although TBM and HDAC maybe contributing to the patients dyspnea it is not the sole etiology and the severe asthma or COPD should be treated aggressively.

Chest CT scanning with dynamic expiratory imaging is a valuable test. The Chest CT will identify the severity of TBM and HDAC when the patient forcefully exhales and the CT scan captures the trachea and bronchi in the expiratory state[1-2]. The CT scan will identify whether TBM or HDAC is the primary clinical entity. And finally, the CT scan may identify a specific clinical entity responsible for TBM or HDAC, which may suggest a specific clinical treatment.

Bronchoscopy is an invaluable tool in the diagnostic work up of TBM and HDAC. Bronchoscopy allows for the real time evaluation of the tracheal and bronchial tree with tidal respirations and with forced expiratory maneuvers. Bronchoscopy with airway exam is considered the gold standard to diagnose TBM and HDAC[1]. It is important to note that the intent is not to elicit coughing during the bronchoscopy and to not overly sedate the patient. Coughing can elicit violent movements of the airway and may result in airway collapse or near complete narrowing in individuals with mild TBM or HDAC and in some patients with normal airways. Patients should not be overly sedated, as they must be able to follow commands and to inhale and exhale on command. The intent of performing the bronchoscopy is to visualize the airway with the patient exhaling forcefully. The bronchoscopy will allow for grading the severity of airway narrowing as well as the location of the narrowed airway segments. In selected cases bronchoscopy will also allow for endobronchial biopsies.

In patients who have complaints of gastroesophageal reflux or who have suspicious findings on chest imaging a pH or impedance probe test is indicated. The test will identify the presence of acid and non-acid reflux into the proximal aero-digestive tract. The cessation of aspiration of acid and non-acid gastric material can decrease airway inflammation and potentially reverse the inciting injury causing TBM and HDAC[9]. Although there may be non-surgical means to reduce aspiration a surgical procedure (such as Nissen fundoplication) may be indicated[9].

Classification

It is important to identify the location, type, and severity of TBM and HDAC as it will guide therapeutic decisions. TBM and HDAC that are located distally in the airway is not amenable to airway stenting or surgical treatment. Distal TBM and HDAC can only be treated with medical management. TBM and HDAC that involves the trachea and/or the proximal main stem bronchi may be amenable to airway stenting and surgical intervention.

The type of airway luminal narrowing may provide insight into the causative etiology. Saber sheath tracheal narrowing is primarily associated with chronic obstructive pulmonary disease[2]. Concentric airway narrowing or severe airway flattening can be seen with relapsing polychondritis or in situations where airway inflammation has decreased the structural integrity of the cartilaginous structures of the trachea and main stem bronchi[8]. (Figure I)

Grading the severity of the TBM and HDAC guides the intensity of the treatment that patients receive. Patients with mild (luminal narrowing of 50% to 70%) and moderate (luminal narrowing of 70% to 90%) TBM and HDAC are treated with medical management with a focus on the primary inciting etiology. Patients with severe (luminal narrowing greater than 90%) and significant symptoms are treated aggressively. Patients with severe TBM and HDAC will receive aggressive treatment of their primary inciting etiology as well as consideration for stent placement and a surgical airway intervention[1-2].

Treatment

Treatment of TBM and HDAC is dependent on symptoms and severity. In patients with severe TBM or HDAC and no clinical symptoms treatment is not warranted. Patients with mild TBM or HDAC and moderate to severe respiratory symptoms another etiology behind their clinical complaints should be sought. Patients with moderate to severe TBM or HDAC and clinical symptoms consistent with TBM or HDAC, aggressive therapy of their TBM or HDAC should be pursued.

Treatment should always include a component of medical therapy[1-2]. Dependent on the clinical situation and the suspected (or diagnosed) clinical etiology of TBM or HDAC medical treatment may be sufficient[1-2]. Patients whose TBM or HDAC is due to severe aspiration or severe recurrent infections appropriate management may sufficiently resolve their symptoms[1-2,8].

Weight management must be included in any treatment regimen for TBM or HDAC. As the abdomen and chest wall become more obese, the configuration of the chest cavity changes with elevation of the diaphragm and the chest wall moving down. The changes in the diaphragm and the chest wall serve to foreshorten the trachea and can decrease the rigidity of the trachea. The decrease in structural integrity of the trachea can allow for a decrease in the amount of pressure required to collapse the trachea and lead to severe tracheal narrowing. Weight loss can reverse the changes to the chest wall and serve as an effective treatment.

Patients with coexistent COPD or Asthma should be maximally treated[1-2]. If maximal treatment of their COPD or Asthma alleviates their complaints, no further airway interventions should be explored. In addition exercise programs should be encouraged as well as proper breathing techniques.

Patients with coexistent obstructive sleep apnea and severe TBM or HDAC a therapeutic trial of non-invasive positive pressure ventilation (NIPPV) trial should be considered. Trials with NIPPV can effectively treat OSA related to TBM or HDAC as well as improve mucociliary clearance[2]. NIPPV serves to pneumatically stent the airway. Pneumatic airway stenting may serve to permit airway clearance and decrease airway inflammation thus treating TBM and HDAC[2,10].

Patients with relapsing chondritis or other collagen vascular associated disease affecting airway integrity may be effectively treated with immune suppression[8]. Aggressive immune suppression may decrease cartilage inflammation and increase airway integrity thereby stabilizing the airway.

Patients who are aggressively treated with medical management and continue to have symptoms due to TBM or HDAC should be considered for definitive surgical treatment. Patients with TBM or HDAC should have a stent trial to document improvement in their symptomatology and objective improvement in their functional status[11]. Generally

an inverted “Y”, silicone stent is deployed via a rigid bronchoscopy (details of rigid bronchoscopy and stenting are covered in their respective chapters in this edition of the journal). These stents are usually left in the tracheobronchial tree for a couple of weeks. Improvement or lack there off is tested with a repeat six-minute walk test, change in ability to perform day-to-day activities or improvement in pulmonary physiology on pulmonary function testing. Long term use of airway stents should be avoided due to the incidence of complications including inspissation of secretions in the stent leading to its blockage and granulation[12]. Patients who fail to improve with a therapeutic stent trial should not be considered for surgical intervention. Surgical intervention (tracheobronchoplasty) involves suturing the posterior membrane to a Marlex mesh to increase the stiffness and rigidity of the posterior membrane[11]. This surgical procedure requires a right thoracotomy and a lengthy operation. Only select centers and surgeons in the USA are experienced in this labor-intensive procedure. In highly selected patients surgical intervention improves respiratory symptoms, health-related quality of life, and functional status[11].

Laser application has been used to treat HDAC[2]. Patients with HDAC with focal areas of the membranous portion of the trachea or bronchi can be treated with topical laser therapy[2]. The intent is to induce scarring that contracts the posterior membrane and functionally stiffens the posterior membrane preventing collapse[2,13].

Future developments may include biogenic stents that slowly become integrated into the airway wall. Biogenic stents that can be placed in a minimally invasive fashion and that allow for normal mucociliary clearance and that enhance airway wall integrity would be very desirable and may have increased utility.

Conclusion

TBM and HDAC can be indistinguishable at the time of presentation. TBM and HDAC differ in the airway anatomy affected. The prevalence of TBM and HDAC in the general population is not known however the incidence in selected populations can be as high as 25%. Patients who have significant respiratory complaints without a defined respiratory diagnosis should be assessed for TBM or HDAC. Testing to diagnosis TBM or HDAC should include chest CT, PFT’s, and a careful clinical evaluation with additional testing guided by clinical findings. Management of patients with TBM and HDAC should include aggressive medical therapy as indicated. Only patients who fail to improve with medical management should proceed to a stent trial. Airway stent trial is to select patients who will benefit from tracheobronchoplasty. Patients who have symptomatic and functional improvement should proceed with trachebronchoplasty. Airway stenting is not a viable long term option due to airway stent complications.

Disclaimers:

Joseph C. Seaman MD – None
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