Recognizing tracheobronchomalacia

August 01, 2006
By Septimiu D. Murgu, MD [1] and Henri G. Colt, MD [2]

Abstract: Tracheobronchomalacia is a form of expiratory central airway collapse characterized by softening of the airway wall cartilaginous structures. Symptoms often mimic asthma and chronic obstructive pulmonary disease. Pulmonary function test results may suggest a diagnosis, but findings are neither sensitive nor specific. Bronchoscopy and novel dynamic radiographic studies contribute to the diagnosis and help differentiate true malacia from other forms of expiratory central airway collapse. Treatment options include medication; noninvasive ventilatory support; interventional bronchoscopy with airway stent insertion; and open surgical procedures, such as tracheostomy, tracheal resection, and tracheoplasty. (J Respir Dis. 2006;27(8):327-335)

Tracheobronchomalacia (TBM) is a disease of the central airways characterized by weakness of the tracheal and bronchial walls caused by the softening of supporting cartilages. As a result, the airways lose their stiffness and the airway walls move closer together, especially during expiration.

The congenital form of TBM may be associated with other conditions, some of which allow survival of affected children into adulthood. Congenital syndromes and anomalies associated with TBM include Ehlers-Danlos syndrome, trisomy 21 syndrome, tracheoesophageal fistula, double aortic arch, and pectus excavatum. Pediatric TBM has been recently reviewed. The adult forms of TBM are either idiopathic or acquired during the course of other illnesses. TBM has also been described as tracheobronchial collapse, expiratory tracheobronchial collapse, expiratory tracheobronchial stenosis, and tracheobronchial dyskinesia. More recently, it seems that true malacia should be regarded as a form of expiratory central airway collapse. The incidence of TBM is unknown; however, it appears to be more common in middle-aged and elderly men who are smokers than in other populations.

In this article, we review the signs and symptoms of TBM and its known causes, and we describe diagnosis and treatment.

PATHOGENESIS

The malacic airway has a crescent-shaped or "saber-sheath" appearance on bronchoscopic or radiographic examination, depending on whether the anterior or lateral walls of the airway, respectively, are weakened (Figure 1). The abnormality is circumferential when a combination of crescent-type and saber-sheath-type narrowing occurs. This abnormality is often seen in patients with relapsing polychondritis. The normal tracheal cartilage-to-soft tissue ratio is approximately 4.5:1. In patients with TBM, however, this ratio is often as low as 2:1. The cartilaginous rings may be destroyed or thinned out, limiting airflow. Atrophy of elastic fibers in the air-ways of these patients has also been described, but this probably characterizes the entity referred to as excessive dynamic airway collapse. Although excessive dynamic airway collapse is seen mainly in patients who have emphysema and asthma, it can also be idiopathic.

TBM is a form of dynamic airway obstruction that may cause hyperinflation and air trapping. During normal expiration, an increase in intrathoracic pressure causes intrathoracic airway narrowing, which is counteracted by tracheobronchial rigidities, elevated intraluminal pressures, and structural attachments. In the malacic intrathoracic airway, however, expiratory collapse occurs when intrathoracic pressure exceeds intratracheal pressure. This event limits airflow in patients who have malacia, and it may eventually contribute to dyspnea, difficulty in clearing secretions, recurrent infections, and even respiratory failure.

ETIOLOGY

The causes of TBM include the following:

- **Indwelling tracheotomy and endotracheal intubation with inflatable cuffs** are considered to be common causes of acquired TBM, most likely resulting from pressure necrosis, decreased blood supply, infection, and friction of tubes on the airway mucosa. The malacic areas are focal and are usually seen at the site of the inflatable cuff, adjacent to the tracheostomy stoma, or at the point of impingement of the tip of the tracheostomy or endotracheal tube on the tracheal wall. Long-term ventilation has been described in patients with Duchenne dystrophy who have had many years of positive pressure ventilation via tracheostomy. It is not clear whether the myopathic
involvement of the tracheal muscularis also contributes to the weakness of the wall. Malacia has also been described in patients who have had long-term tracheostomies.\textsuperscript{13}

Closed chest trauma may cause unrecognized tracheal fractures, which can result in malacia. This is most likely the result of impairment in the blood supply.\textsuperscript{10}

Chronic irritation of the airways may weaken the airway wall. It can be the result of cigarette smoking or air pollution exposure, which are recognized risk factors for TBM.\textsuperscript{14}

Chronic inflammation can cause progressive atrophy and destruction of the tracheal or bronchial cartilages. This is most significant in patients with relapsing polychondritis, more than half of whom show involvement of the tracheobronchial tree.\textsuperscript{15,16}

Malignancy may cause focal malacia, such as that seen in tracheal or bronchial cancers or in extraluminal tumors that extend into and destroy the airway walls.\textsuperscript{1}

Mechanical anatomic factors, including postpneumonectomy syndrome\textsuperscript{17} and lung and heart-lung transplantation, can lead to malacia. In such cases, airway abnormalities are seen at or below the site of airway anastomoses.\textsuperscript{18}

Infections, such as tuberculosis, may cause a progressive destruction or atrophy of cartilaginous rings.\textsuperscript{19}

Chronic compression of the tracheobronchial tree, as seen in substernal goiters and vascular anomalies, can lead to malacia.\textsuperscript{20,21}

Congenital tracheobronchomegaly (also known as Mounier-Kuhn syndrome) is a congenital abnormality of the tracheobronchial tree characterized by atrophy or absence of longitudinal elastic fibers and thinning of the muscularis mucosa.\textsuperscript{22} This most likely causes excessive dynamic airway collapse rather than malacia.

Ehlers-Danlos syndrome has been described as one cause of laryngotracheomalacia. It is not known whether this syndrome results from compression by dilated aortic and pulmonary arteries\textsuperscript{23} or from an intrinsic cartilaginous defect of the wall.

Thyroid masses cause prolonged compression of the trachea, especially within the confines of the thoracic inlet. Tracheal cartilaginous ring involvement, most commonly after thyroidectomy or as a result of a tumor, can also cause malacia.\textsuperscript{24,25}

Endobronchial electrosurgery has been reported as a cause of cartilage damage and bronchomalacia in animal models and humans.\textsuperscript{26,27}

Chronic obstructive pulmonary disease (COPD) and asthma can be accompanied by excessive dynamic airway collapse. There have been numerous reports of excessive dynamic airway collapse associated with COPD and asthma, although it was frequently referred to as TBM or tracheobronchial collapse.\textsuperscript{4,5,10} Chronic inflammation probably contributes to the atrophy of the elastic fibers of the membranous portion of the tracheobronchial wall.\textsuperscript{28} Chronic irritation and cough may also weaken the tracheobronchial wall as a result of repeated, excessive pressure changes. 

**DIAGNOSIS**

**Clinical manifestations**

In patients with TBM, dyspnea, cough, difficulty in clearing secretions, recurrent bronchitis, pneumonia, and respiratory failure have been reported.\textsuperscript{1,2,5,7,28} Cough is usually described as seal-like barking, which probably is caused by the vibration of the floppy membranous wall against the anterior airway wall during expiration.\textsuperscript{17}

Wheezing is present in more than half of patients with TBM. Asthma-like exacerbations, characterized by wheezing and dyspnea, are less common and are usually refractory to corticosteroids and bronchodilators.\textsuperscript{28} Most patients have dyspnea on exertion, probably because during exercise, the airway collapse is worsened by increased intrathoracic pressures. Hemoptysis is rare in patients with TBM.\textsuperscript{29} Hypercapnic respiratory failure that requires mechanical ventilation or presents as an inability to wean off mechanical ventilation has been reported.\textsuperscript{30,31}

A diagnosis of TBM should be considered in patients with obstructive ventilatory impairment who do not respond to conventional treatment with bronchodilators or inhaled corticosteroids. Any patient at risk for TBM should be evaluated if they have chronic respiratory symptoms. TBM should also be in the differential diagnosis of unexplained hypercapnic respiratory failure, especially in cases of failed extubation after successful weaning trials.

TBM is a progressive disease in adults\textsuperscript{2,28} and in severe cases can be fatal; however, no reliable mortality data are available.

**Pulmonary function testing**

Few studies have rigorously reported results of pulmonary function testing in patients with malacia,\textsuperscript{4,5,32,33} and differences in disease definition account for considerable variability among those studies that do describe findings.\textsuperscript{5,33} The expiratory spirometry may reveal a “typical notch” on the volume-time curve,\textsuperscript{33,34} which might reflect a sudden diminution of flow at the beginning of expiration when the airway collapses. This spirogram is characterized by an initial phase in which a
Recognizing tracheobronchomalacia
Published on Psychiatric Times
(http://www.psychiatrictimes.com)

small volume is rapidly exhaled, followed by an upward deflection and then the continuation of exhalation.\textsuperscript{4,5}

In patients with TBM, the flow-volume loop patterns on a spirogram may suggest compression of the central airways (Figure 2). The maximal flow is reached quickly after expiration of a small volume of air. Following the maximal flow, there is a significant decrease in flow, although only a small volume is exhaled. Then, the flow rate falls very little during the remainder of expiration; this phase is responsible for the long plateau of the flow-volume loop. Flow oscillations have also been described; they have a “saw-tooth” appearance, which is a reproducible sequence of alternating decelerations and accelerations of flow.\textsuperscript{32,34}\textbf{Dynamic imaging studies}

Standard chest radiography and single-slice CT scanning are performed at end-inspiration and do not permit the precise assessment of airway collapse that is required to diagnose true malacia. Traditional fluoroscopy offers poor-quality display of anatomic detail of the paratracheal structures and fails to fully visualize the airways in obese patients.\textsuperscript{35} Despite their limitations, these techniques may help diagnose airway collapse. They are relatively inexpensive and might therefore be used when more sensitive and specific diagnostic tests are not readily available. However, dynamic CT allows volumetric acquisition of data both at end-inspiration and during dynamic expiration (Figure 3). Many investigators define malacia as a reduction in airway caliber of 50\% or more between inspiration and expiration,\textsuperscript{35-38} but other criteria have been proposed.\textsuperscript{39} Dynamic CT correlates well with bronchoscopy findings,\textsuperscript{37} reveals air trapping, offers good display of anatomic detail of the airway and adjacent structures, and allows for objective interpretation and measurement of the degree of collapse.\textsuperscript{35} Cine MRI appears to be sensitive in the diagnosis of malacia, but clinical experience is still limited.\textsuperscript{40,41}\textbf{Dynamic and quantitative bronchoscopy}

Flexible bronchoscopy is considered by some to be the gold standard for diagnosing TBM.\textsuperscript{2} Asking the patient to breathe deeply, cough, and exhale can elicit the collapsibility of the airways. Dynamic bronchoscopy allows the examiner to identify changes in airway caliber; note the extent of airway collapse; and classify airway narrowing as being of the crescent, saber-sheath, or circumferential type.\textsuperscript{3} Morphometric bronchoscopy,\textsuperscript{42-45} a technique by which maximal and minimal cross-sectional areas are measured, may have a future diagnostic role. Endobronchial ultrasonography shows distinct hypoechoic and hyperechoic layers as laminar structures of the airway walls.\textsuperscript{46} This procedure can also identify destroyed cartilage.\textsuperscript{47}\textbf{Differential diagnosis}

The differential diagnosis of TBM includes excessive dynamic airway collapse and saber-sheath trachea. Excessive dynamic airway collapse is a clinical finding that is described as the exaggerated inward bulging of the posterior membrane. This causes a reduction of 50\% or more in the airway lumen area, as documented on fluoroscopy, dynamic CT, cine MRI, or flexible bronchoscopy during exhalation.\textsuperscript{1,9} Excessive dynamic airway collapse is often present in patients with asthma and emphysema. The presentation, which can include dyspnea, cough, wheezing, and recurrent respiratory infections, may be indistinguishable from that of true malacia. The expiratory collapse of the posterior membrane can be observed in the presence or absence of cartilaginous damage. Saber-sheath trachea is a fixed narrowing described in up to 5\% of older men with COPD. It is characterized by a fixed reduction of the transverse diameter of the intrathoracic portion of the trachea; the internal transverse diameter is not more than two thirds of the size of the internal anteroposterior diameter at the position 1 cm above the aortic arch.\textsuperscript{48} Instead of a weakening of the cartilaginous rings, histopathology reveals thickened and calcified tracheal rings.\textsuperscript{48} This condition is different from the saber-sheath form of malacia, in which the expiratory collapse of the lateral airway is dynamic and is caused by cartilage damage.\textsuperscript{49} TBM is an important addition to the differential diagnosis of difficult-to-control asthma, COPD, and breathlessness. Signs and symptoms of TBM often imitate those of asthma and COPD. In some cases, TBM is accurately diagnosed only after the patient has been treated for months or years for a refractory obstructive ventilatory disorder.\textbf{Classification}

Proposed classifications for true malacia have not been universally adopted because of a lack of clarity in definitions, measurement criteria, and terminology.\textsuperscript{10,14,28,50,51} In the proposed "FEMOS" classification system, the degree of expiratory central airway collapse is identified according to functional class, extent, morphologic type, origin, and severity (degree of airway narrowing) (Table).\textsuperscript{9} This multidimensional classification system requires validation.\textbf{TREATMENT}

Therapy depends on the severity of symptoms, the degree and extent of airway collapse, and the underlying cause of TBM.\textsuperscript{1}\textbf{Conservative management}

If permitted by the patient’s clinical status, treatment of the underlying condition should be
Recognizing tracheobronchomalacia

Published on Psychiatric Times
(http://www.psychiatrictimes.com)

optimized before considering invasive therapies. Drug therapy should be used whenever possible before proceeding to more aggressive interventions. This is particularly the case for relapsing polychondritis, for which the main therapy consists of NSAIDs for mild disease and high-dose corticosteroids and various immunosuppressants for more severe forms. Corticosteroids are often effective in treating chondritis; they decrease the frequency and severity of recurrence. They do not, however, stop the late loss of cartilaginous support and, in fact, may not influence survival. Noninvasive positive pressure ventilation can be used to maintain airway patency, facilitate secretion drainage, and improve expiratory flow. The addition of nasal continuous positive airway pressure (CPAP) improves spirometry values, sputum production, atelectasis, and exercise tolerance. Although large controlled studies are needed to confirm these findings, it seems that nocturnal and intermittent daytime nasal CPAP benefits patients with TBM and can be used as adjunctive therapy. Although various levels of CPAP were applied, 10 cm H2O of CPAP appeared optimal for all patients, based on observed plateaus in expiratory airflow, near-normalization of airway collapse during active expiration, and degree of patient tolerance and comfort.

**Minimally invasive surgery**

Airway stents can successfully maintain airway patency and result in improved pulmonary function (Figure 4). If symptoms do not improve, stent removal is probably required to avoid stent-related complications, including migration, obstruction by mucus or granulation tissue, infection, fracture, and airway perforation. Close follow-up is warranted and any recurrence of symptoms of TBM should prompt immediate bronchoscopy.

**Open surgery**

Tracheostomy may stent the malacic airway and provides invasive ventilatory support when necessary. Tracheostomy, however, can be complicated by secondary tracheomalacia and stenosis at the stoma site and should probably not be considered as a first-line treatment in elective cases. Tracheal resection has been proposed for focal tracheomalacia; it has been shown to have good outcome and low mortality in experienced centers.

Airway splinting via tracheoplasty has been used to consolidate and reshape the airway wall. This technique allows reinforcement of the membranous portion of the trachea in crescent-type malacia with outcomes that appear favorable in uncontrolled studies.

Other techniques that have been reported include tying the posterior wall of the trachea with bone chips, fascia grafts, or plastic prostheses; performing autologous cartilage grafts to support the tracheal wall; suturing the trachea to dura mater grafts; and implanting biocompatible ceramic rings.

**References: REFERENCES**

12. Baydur A, Kanel G. Tracheobronchomalacia and tracheal hemorrhage in patients with Duchenne...
Recognizing tracheobronchomalacia
Published on Psychiatric Times
(http://www.psychiatrictimes.com)


Source URL: [http://www.psychiatrictimes.com/articles/recognizing-tracheobronchomalacia](http://www.psychiatrictimes.com/articles/recognizing-tracheobronchomalacia)

Links:
[1] [http://www.psychiatrictimes.com/authors/septimiu-d-murgu-md](http://www.psychiatrictimes.com/authors/septimiu-d-murgu-md)
[2] [http://www.psychiatrictimes.com/authors/henri-g-colt-md](http://www.psychiatrictimes.com/authors/henri-g-colt-md)