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Tracheobronchomalacia in the Adult: Is Imaging Helpful?

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Abstract

Purpose of Review The aim of this article is to: 1) Review the imaging features of tracheobronchomalacia and excessive dynamic airway collapse 2) To highlight the recent advances in imaging of the central airways.

Recent Findings Imaging of the central airways requires dedicated techniques optimized to evaluate the transient collapse of the central airways either due to excessive movement of the posterior membrane and/or secondary to weakness of the tracheal cartilages. Routine imaging of the chest is limited in its ability to demonstrate the true extent of collapsibility of the airways. The more recently introduced imaging protocols utilize dynamic acquisition during active expiration which demonstrate the transient but true extent of the airway collapse. CT also has potential applications in treatment in the form of 3D-printed splints.

Summary Dedicated imaging protocols introduced with recent advances in CT technology allow diagnosis of tracheobronchomalacia (TBM) noninvasively. However, there is a current lack of clear threshold for diagnosis which has led to a wide range of reported prevalence of TBM and excessive dynamic airway collapse.

Keywords Tracheobronchomalacia · Dynamic airway collapse · Multidetector CT imaging

Introduction

Tracheobronchomalacia (TBM) and excessive dynamic airway collapse (EDAC) are diseases that are characterized by exaggerated narrowing of central airways on expiration. They can occur independently or together and have different pathophysiologic causes. TBM specifically refers to weakness of tracheobronchial walls secondary to abnormality of the supporting cartilage, whereas EDAC relates to excessive forward displacement of the posterior membrane during expiratory phase (Fig. 1a, b) [1•]. When EDAC occurs in isolation, it represents a lax posterior membrane with structurally intact cartilage. Multidetector computed tomography (MDCT) is increasingly being used in the evaluation of TBM/EDAC, and therefore, it is important to be aware of its strengths, limitations, current controversies in diagnosis, and recent advancements such as 3D printing [2, 3].

This article will focus on the following aspects:

- Discuss the types and causes of TBM
- Role of state-of-the-art imaging in the evaluation of TBM and EDAC
- Critically evaluate relevant literature to emphasize key findings, controversies surrounding diagnosis, and management

Classification

TBM is increasingly being recognized as one of the causes of chronic respiratory symptoms. The recent emphasis on TBM is partly due to advancements in CT technology which allow faster and volumetric acquisition of the airways during inspiration and during active expiration [4]. The accuracy of dynamic CT of airways has been shown to be similar to bronchoscopy as the previously recognized reference standard [5].

Classification of TBM is important for therapeutic decision-making and management. TBM is commonly classified based on extent, distribution, and morphology [6]. The weakness of airway cartilage may be diffuse or segmental

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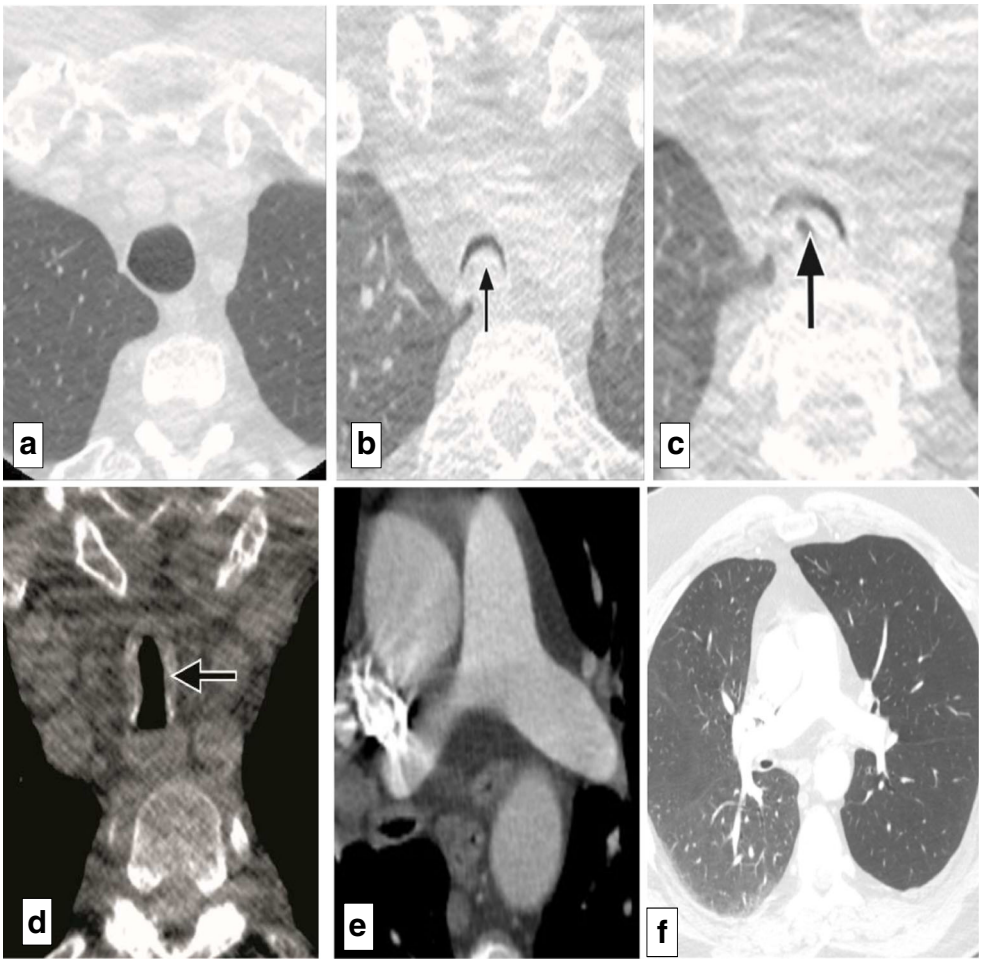

Fig. 1 Normal configuration of the trachea on inspiration (a). Excessive dynamic airway collapse (EDAC) characterized by marked inward bowing (arrow) of the posterior membrane during dynamic expiration with normal integrity of C cartilages (b). Crescent type tracheomalacia with abnormal flattened configuration of the anterior tracheal cartilage in association with exaggerated bowing of the posterior membrane leading to a markedly collapsed tracheal lumen (arrow) on expiration (c). Saber sheath tracheomalacia with weakening of the lateral aspects of the tracheal cartilage leading to side by side narrowing (d). Circumferential type bronchomalacia with global abnormality of left main stem bronchial cartilage (e) and asymmetric diffuse air trapping in left lung on expiration (f)

involving only portions of the tracheobronchial tree [7]. Isolated tracheal involvement is referred to as tracheomalacia (TM). The term TBM is used when the weakness also involves the main stem bronchi.

Depending on the morphology and the shape of the central airways, TBM has been classified as:

- Crescent type—abnormality of the anterior aspect of the tracheal cartilage leading to narrowing in anteroposterior dimension (Fig. 1c)
- Saber-sheath type—abnormality of the lateral aspect of the tracheal cartilage leading to side-to-side narrowing (Fig. 1d)
- Circumferential-global abnormality of the tracheal cartilage (Fig. 1e, f)

Etiology

TBM can be congenital or acquired, with acquired causes being more common. The various etiologies are summarized in Table 1. Common congenital anomalies associated with TBM include mucopolysaccharidosis, trisomy 9 and 21, tracheoesophageal fistula, and bronchopulmonary dysplasia [8, 9]; Ehlers-Danlos syndrome and Mounier-Kuhn syndrome can also lead to TBM due to atrophy of longitudinal elastic fibers with thinning of muscularis mucosa [10].

Acquired causes can be broadly categorized into traumatic, mechanical, and inflammatory processes. Traumatic causes are by far the most common and are often iatrogenic such as following prolonged intubation, tracheostomy, and in post-operative setting (e.g., after lung transplant) [6, 11].

Mechanical causes include wall weakening due to prolonged compression of the trachea by mass or vascular anomalies. Mass effect from various types of vascular anomalies such as vascular rings may cause secondary tracheomalacia or the presence of associated structural defects of the tracheobronchial system [12, 13]. Recognition of TBM is important in these cases as airway narrowing may persist even after surgical resection of the mass or correction of vascular anomaly [8].

Various airway inflammatory processes can be associated with TBM including emphysema and obstructive lung disease. It has been reported that airway collapse

Table 1 Etiologies congenital or acquired of TBM

Congenital	Storage diseases	Mucopolysaccharidosis
	Chromosomal anomalies	Trisomy 21 and 9
	Anomalous anatomy	Bronchopulmonary dysplasia
	Connective tissue diseases	Tracheobronchial fistula
		Ehlers-Danlos syndrome
Acquired	Post-traumatic	Prolonged intubation
		Tracheostomy
		Tracheal anastomosis as in lung transplant surgery
	Mechanical	Extrinsic compression from lymph nodes or lung mass
		Vascular anomalies/aneurysms
		Skeletal abnormalities, e.g., pectus excavatum
	Inflammation	Relapsing polychondritis
		Radiation therapy
	Emphysema/COPD	Chronic bronchitis

of > 50% is seen in about 12.7% in patients with COPD and smoking-related lung disease on bronchoscopy [14, 15]. Relapsing polychondritis is another inflammatory condition that causes destruction and weakness of the cartilaginous structures of the external ear, nose, and tracheobronchial tree resulting in an excessive collapse of the airways [16]. It has been reported that while 56% of cases of relapsing polychondritis involve the tracheobronchial tree, only 14% of patients present with respiratory symptoms [17, 18].

True prevalence of TBM is difficult to ascertain due to the uncertainty of diagnostic threshold to define TBM. Gangadharan et al. [19] report that 13% of emphysema patients have TBM using the standard criteria of 50% collapse. However, when the threshold is raised to 70%, only 5% of patients would fit the criteria for TBM. In our institution, we use > 70% decrease in the luminal cross-sectional area on dynamic expiration as the diagnostic criteria for clinical tracheobronchomalacia.

Clinical Presentation

Mild forms of TBM may be asymptomatic. The severity of symptoms and signs is related to the degree and extent of TBM [2, 4, 20]. The symptoms, when present, are at best nonspecific and mostly attributed to coexisting conditions such as COPD, asthma, smoking, and bronchogenic carcinoma. If clinically significant, TBM may present with various manifestations including dyspnea, cough (sometimes productive), hemoptysis, wheezing, and stridor. The cough usually has a barking character and may be associated with choking sensation. Recurrent pulmonary infections are often reported [21]. In patients on mechanical ventilation, TBM may manifest as failure to wean [22].

Diagnosis

Diagnosis of TBM is based on clinical findings and confirmed with diagnostic tests. Bronchoscopic visualization of dynamic airway collapse has been the reference standard for the diagnosis of TBM [23]. Multidetector computed tomography (MDCT) technique using dynamic expiratory CT during forced expiration is now being used as a noninvasive test to confirm the diagnosis of TBM with accuracy ranging from 93 to 97% [1•, 24–26].

CT Imaging Technique

The routine high-resolution computed tomography (HRCT) with end-inspiratory/end expiratory scans can miss the transient collapse of the central airways and is not preferred for TBM.

In adults, volumetric paired end-inspiratory/dynamic expiratory MDCT is the most reliable imaging technique [27]. This involves imaging at end-inspiration as with routine HRCT, followed by imaging during forceful exhalation, called the dynamic expiratory component. The area of coverage extends from the larynx down to the main bronchi. For an optimal depiction of airways, thin slice collimation of 0.6 to 1.25 mm with overlapping reconstruction intervals of 50% is preferred to obtain the isotropic dataset. This prevents stair-step artifacts on multiplanar reconstructions (MPRs) (Fig. 2) [28].

The volumetric end-inspiratory component provides the baseline anatomic detail of the central airways during full inspiration, depicting the size and shape of the airway lumen, any intrinsic or extrinsic masses in relation to the airways, and also the distribution of the airway wall thickening if present (Fig. 3). The relationship of the airways to the adjacent mediastinal and hilar structures is also assessed on these images [4].

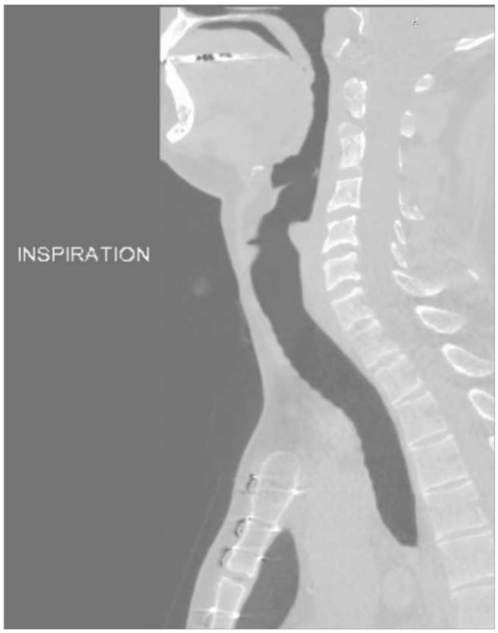

Fig. 2 Sagittal reconstruction of the trachea derived from volumetric isotropic voxel dataset demonstrating good image quality with no stair-step artifacts

For the dynamic expiratory component of the study, optimal breathing instructions are of paramount importance. The patients are instructed to take in a deep breath and exhale forcefully during image acquisition [29]. In our experience, optimal coaching and practicing dynamic expiratory maneuvers prior to the actual scan are essential to avoid errors and misdiagnosis [30]. Table 2 summarizes our institutional protocol for imaging of central airways using dynamic expiratory scanning.

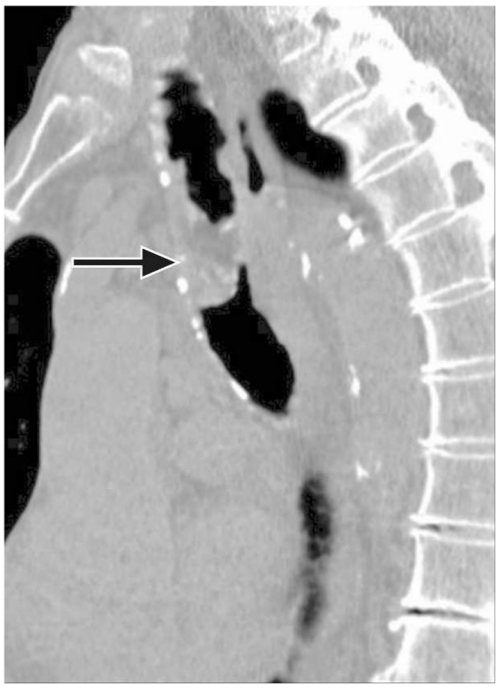

Fig. 3 Sagittal reconstruction of a CT data set showing extent of the tracheal mass (arrow)

Table 2 Institutional HRCT protocol for imaging of central airways

1. Scouts: AP and lateral	
2. Supine inspiration: start, 7 cm below the carina; end, 1 cm above the epiglottis	
Scan type	Helical
Gantry rotation time	0.5 s
Detector coverage	40 mm
Slice thickness	0.625 mm
Interval	0.625 mm
Pitch	1.375:1
KVP	120
Auto mA (min/max)	80/350
DFOV	22 cm
Algorithm	Standard
WW/WL	500/50
3. SUPINE "dynamic" expiration: start, 7 cm below the carina; end, 1 cm above the epiglottis	
Breathing instruction: Start scan at the start of the forceful expiration part of the breathing instructions: "*Take in a breath, let your breath out. Take in another breath and now blow it out as hard as you can*"	
Scan type	Helical
Gantry rotation time	0.5 s
Detector coverage	40 mm
Slice thickness	0.625 mm
Interval	0.6.5 mm
Pitch	1.375:1
KVP	120
Auto mA (min/max)	80
DFOV	22 cm
Algorithm	Standard
WW/WL	500/50

Another technique using cine CT during active coughing maneuver can also be performed on a MDCT scanner with 64 or more rows [20]. With the availability of 320-row MDCT, coverage of the entire tracheobronchial tree becomes possible in most children and adults in one gantry rotation. Intravenous contrast is generally not required for assessment of TBM unless an intrinsic or an extrinsic mass compressing or invading the airways is suspected.

While MDCT is currently the most widely used imaging modality for TBM, more recently, MRI is being studied as another tool to evaluate the dynamic airway collapse without the penalty of radiation exposure. MRI has been shown to be feasible and can be acquired as static 3D images during inspiration and end expiration as well as cine MRI during dynamic breathing [31]. In this study, MRI was proven to be technically feasible though dynamic cine MRI demonstrated greater collapse compared with

MDCT. Larger studies are needed to validate and define the role of MRI in TBM diagnosis and management.

Post Processing and Image Interpretation

Volumetric acquisition permits the generation of high-quality three-dimensional and multiplanar reconstructions for both the end-inspiratory and dynamic expiratory phases which help in diagnosis and surgical planning [32]. MPRs are two-dimensional reformations of the anatomy generated from the volumetric dataset and can be displayed in sagittal and coronal planes or in a curved format along the long axis of the airways [33]. In addition, external 3D rendering of the airways can be generated which allows evaluation of the relationship of airways with adjacent structures. Internal 3D rendering or "virtual bronchoscopy" is a valuable tool which enables navigation through the lumen of the central airways similar to conventional bronchoscopy (Fig. 4) [34]. Several authors have shown the utility of this tool in the rapid and precise assessment of the extent of airway stenosis, in guiding transbronchial biopsy procedures and evaluating endobronchial lesions [35–37].

The axial end-inspiratory images provide excellent anatomic information of the airways and adjacent structures and also serve as a point of reference for the dynamic expiratory images to evaluate airway collapse. However, the axial images can be limited in assessing the complex tubular morphology of the airways. It is therefore helpful to use MPRs and 3D image reconstructions in addition to the axial dataset to look for subtle airway stenosis and to establish the true craniocaudal extent of the disease [38]. This can also help in meaningful communication of the results to the managing clinical teams who can then plan appropriate therapy [39].

The dynamic expiratory images are evaluated for the degree of airway collapse during forced expiration. Notably, the diameter of the central airways changes during normal respiration. The airways dilate slightly during inspiration and become narrow during expiration, mostly due to in-bowing of the posterior membrane. However, in TBM, the weakening of the cartilaginous walls deforms the lumen of the airways during expiration and exaggerates the degree of collapse. When EDAC occurs in isolation, there is excessive in-bowing of the posterior tracheal membrane with intact cartilaginous integrity.

Diagnostic Criteria for TBM and EDAC The degree of luminal narrowing can be assessed visually or more accurately by electronic tracing tools by calculating the cross-sectional area of the lumen at the corresponding levels on end-inspiratory and dynamic expiratory scans [20]. It has been proposed that > 50% narrowing of the lumen during expiration is necessary for the diagnosis of TBM and EDAC [40]; however, some studies have demonstrated that > 50% decrease in the cross-sectional area of the tracheal lumen can be seen in a substantial percentage (up to 78%) of normal healthy individuals [41]. Hence, diagnosis requires clinical correlation and cannot be made solely on the basis of meeting a CT threshold for collapsibility. Because of overlap with normal individuals, a > 70% narrowing of the lumen has been more recently proposed as the diagnostic criteria by some authors (Fig. 5) [5, 27]. This current lack of clear threshold for diagnosis has led to wide a range of reported prevalence of TBM and EDAC. In our practice, if the cross-sectional area of the airway lumen decreases by > 70% on dynamic expiratory imaging, we raise the possibility of excessive airway collapse and request correlation with clinical symptoms as well as any risk factors for TBM (such as prior intubation and polychondritis). Pulmonary function tests can also be performed, though these are neither sensitive nor specific. The term severe airway collapse is typically reserved for 90% or greater expiratory narrowing [42].

Radiation Dose

Radiation dose is a concern given a patient is scanned twice, once during end-inspiration and then during forced exhalation. However, the radiation dose can be significantly reduced by

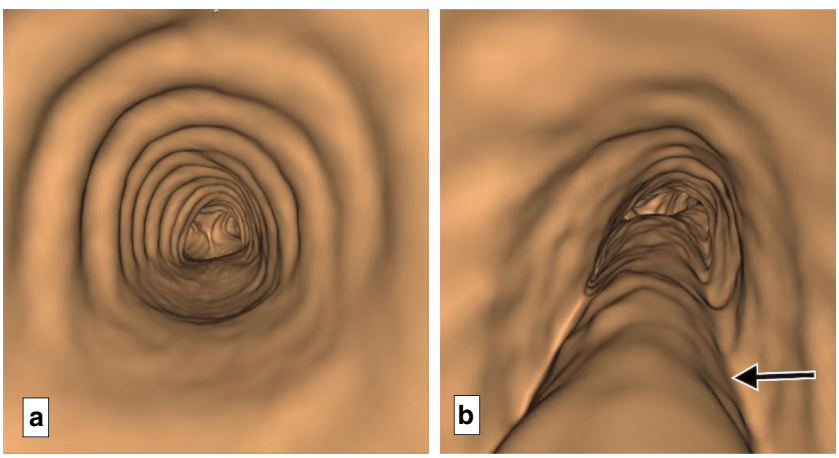

Fig. 4 Virtual bronchoscopy showing normal lumen of trachea in inspiration (a) and excessive bulging of posterior membrane (arrow) during dynamic expiratory imaging in EDAC (b)

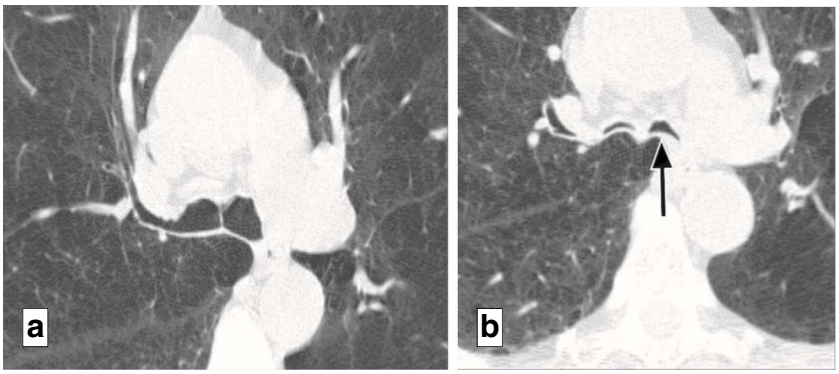

Fig. 5 Axial CT image at the level of carina during inspiration shows normal luminal diameter of central airways (a). Axial CT image at the same level during dynamic expiration (b) shows marked collapse of the central airways (arrow) which is greater than 70%, fulfilling the criteria for the diagnosis of EDAC

using low-dose (30–40 mA) technique during the dynamic expiratory phase and limiting the coverage to the trachea and central airways. The high contrast between the trachea containing air and the surrounding soft tissues permits low-dose technique without

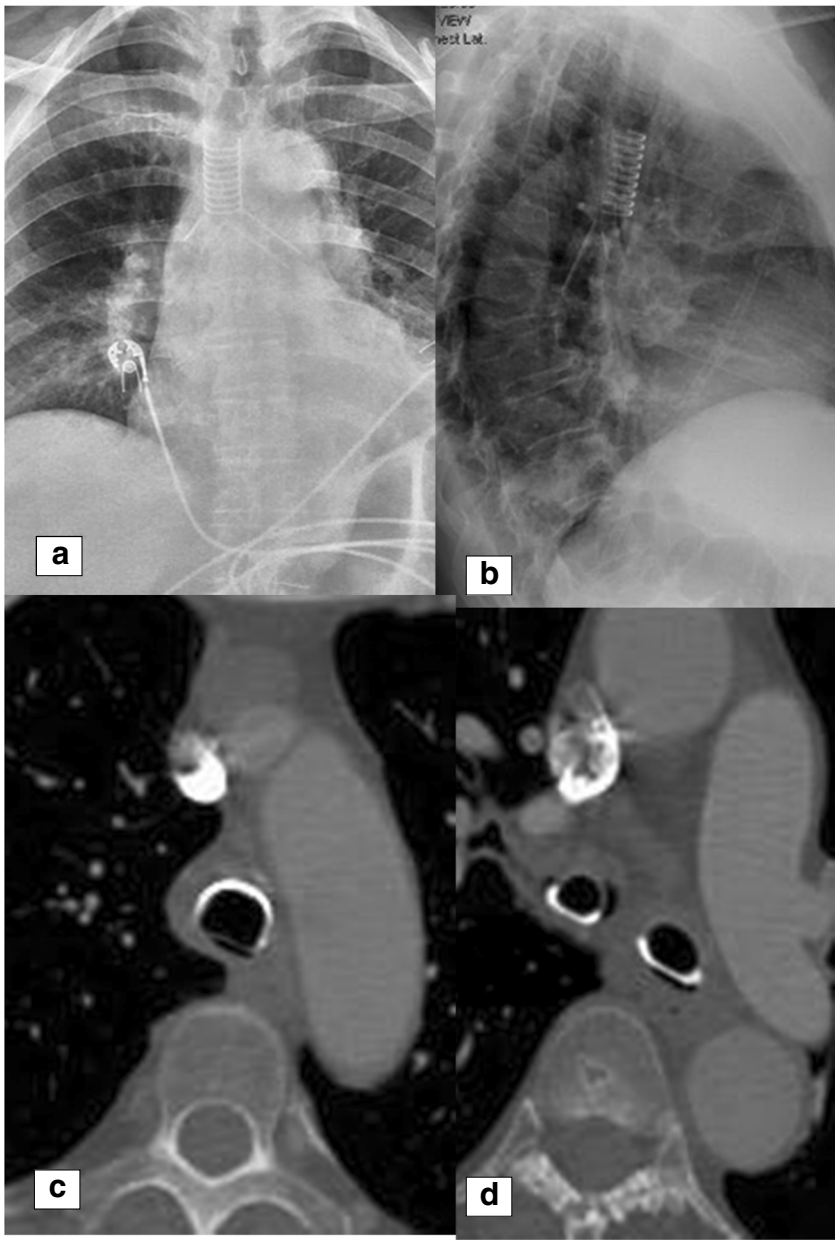

Fig. 6 Frontal (a) and lateral (b) chest radiographs show tracheobronchial Y stent in the trachea and main stem bronchi. Axial CT images at the level of trachea (c) and just below (d) the carina show the patency of the stent. Stents are usually reserved for short term use prior to surgical procedures

limiting the image quality [43]. Dose modulation can be concurrently used to further reduce the radiation dose [4].

Treatment

Various treatment options include medical management, minimally invasive interventions, and surgical approach. The underlying conditions and comorbidities should be treated. Positive-pressure ventilation has been described to act as a pneumatic stent and helps improve drainage of secretions. Various stents (such as silicone stents and self-expanding metallic stents) have also been used and are primarily reserved for short term trial before surgical correction (Fig. 6). This helps to determine what component of patient symptomatology is attributable to airway collapse. Long-term use of stents is justifiable only for those unable to undergo surgery. Surgical options include tracheostomy and tracheoplasty (reinforcement of posterior membrane). New treatment options are also being evaluated including customized bioresorbable 3D-printed splints made from CT dataset [44, 45].

On intermediate follow-up scans after tracheobronchoplasty, CT has shown improvement in expiratory tracheal collapsibility. While the collapsibility increases on long-term follow-up, it still remains significantly improved compared with preoperative baseline [46•].

Summary

Evaluation of TBM is difficult on conventional chest CT as it requires demonstration of transient airway collapse during active breathing. Dedicated imaging protocols have therefore been developed which are continuously evolving and being refined with recent advances in CT technology. Newer scanners with faster scan times and larger volumes of acquisition in a single gantry rotation allow rapid, noninvasive, and accurate assessment of central airways during dynamic expiration to catch the transient changes in airway caliber. CT not only plays an important role in the diagnosis of this entity, but also has potential applications in treatment in the form of 3D-printed splints. Diagnosis remains challenging on imaging findings alone given overlap with the normal population. Hence, correlation with clinical status and multidisciplinary collaboration is essential for the diagnosis of clinically significant TBM. Cine MRI is a potential alternative to MDCT for the diagnosis of central airway disease; however, more studies with larger sample sizes are needed to establish its role.

Compliance with Ethical Standards

Conflict of Interest Aamer R Chughtai and Prachi P Agarwal declare no conflict of interest.

Human and Animal Rights and Informed Consent This article does not contain any studies with human or animal subjects performed by any of the authors.

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