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Diagnosis, Classification, and Management of Pediatric Tracheobronchomalacia
A Review

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IMPORTANCE Tracheobronchomalacia (TBM) describes an increased collapsibility of the trachea and bronchi that is greatest on forced expiration. A broad term, TBM encompasses intrinsic tracheal weakness, some forms of tracheal deformation, and extrinsic compression. Tracheobronchomalacia is the most common congenital tracheal anomaly, affecting 1 in 2100 children. Tracheobronchomalacia is often associated with recurrent and prolonged respiratory tract infections, can lead to chronic lung disease, and can be fatal in its most severe form. Tracheobronchomalacia is often associated with other congenital anomalies and syndromes.

OBSERVATIONS There is a paucity of information on TBM treatment in the modern otolaryngology literature. The primary treatment modalities described include tracheotomy, tracheal stents, and anterior aortopexy. In this review, a new TBM classification scheme and new treatment strategies are introduced to the otolaryngology literature. Diagnosis is made through history and physical examination, dynamic airway computed tomography, and dynamic 3-phase tracheobronchoscopy. Medical management includes nebulizer treatments, minimal use of inhaled corticosteroids, gastroesophageal reflux disease therapy, and continuous positive airway pressure. Surgical techniques, including anterior and posterior tracheobronchopexy and anterior and posterior aortopexy, are described.

CONCLUSIONS AND RELEVANCE Tracheobronchomalacia is an entity of relevance to pediatric otolaryngologists and should be considered as being associated with respiratory distress, stridor, cough, recurrent pneumonia, or feeding difficulties, especially in children with syndromes or other congenital anomalies. A multidisciplinary approach to these patients is essential. A classification scheme facilitates discussion of individual patients among health care professionals and guides appropriate management. Novel surgical approaches for the treatment of TBM, including anterior and posterior tracheopexy and aortopexy, may be considered in management of the treatment of children with symptomatic TBM.

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Video

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Tracheobronchomalacia (TBM) is an imprecise term that is used to describe unilateral airway compression (as seen in innominate artery compression and aberrant subclavian artery compression), circumferential airway compression (as seen in double aortic arch and vascular rings), diffuse or focal cartilage weakness with dynamic collapse of the large airways (as seen in some genetic conditions and tracheal inflammation), and excessive dynamic motion of the posterior tracheal membrane during forced exhalation. Tracheobronchomalacia can occur as 1 form or a combination of 2 or more forms of the above conditions, and different regions of the airway may be affected by different conditions. This complexity makes precise classification of TBM difficult.

Tracheobronchomalacia can be further divided into tracheomalacia (TM), with collapse isolated to the whole or segments of the trachea, and bronchomalacia, a less common entity describing isolated collapse of 1 or both mainstem bronchi.[1,2] The third entity of small airway malacia, involving the lobar bronchi and perhaps smaller airways, is another form that is less well understood and often associated with prematurity, bronchopulmonary dysplasia, and other conditions.

Tracheomalacia was first described in the literature in the 1930s-1940s. In 1952, Holinger et al[3] reported on infants with dynamic collapse of the large airways that resolved with passage of a bronchoscope. Baxter and Dunbar further characterized TM in 1963, describing it as "a condition in which there is a weakness of the tracheal wall due to softening of the supporting cartilage and hypotonia of the myoelastic elements. The result is a trachea which lacks its usual degree of stiffness and the anterior and posterior walls coapt resulting in collapse of the tracheal lumen."[4(p1013)]

In healthy individuals, the tracheal lumen dilates on inspiration due to outward posterior motion of the posterior membrane with

decreasing intrathoracic and mediastinal pressures relative to the intraluminal airway pressures. The tracheal lumen narrows on expiration as the increased intrathoracic and mediastinal pressures relative to the intraluminal airway pressures causes airway compression and the posterior membrane moves inward and anteriorly. The shape and strength of the cartilage and narrow posterior wall and the tone of the pars membrenacea allow the healthy trachea to withstand collapse even with the high intraluminal pressures generated during cough and forced exhalation.[2,5] The healthy trachea will narrow less than 50% on vigorous coughing in adults.[1,2]

In patients with TM, the tracheal cartilage often has an aberrant shape: instead of the normal C shape with a narrow posterior membrane, the cartilage may be more of a U shape with a wider and more mobile posterior membrane, or even a bow shape with a very wide and very mobile posterior membrane. Histopathologic studies of the tracheas of children with TBM/tracheoesophageal fistula (TEF) postmortem found a decrease in the normal 4.5:1 ratio of cartilage to posterior membrane muscle.[6] In most cases, TBM affects the intrathoracic trachea and narrowing and/or collapse of the airway is most severe on forced expiration.[2,7] Historically, a dynamic reduction in the tracheal lumen of 50% has been used as the threshold to diagnosis of TBM, although this reduction must be correlated with symptoms to be considered pathologic.[7]

One common classification system is as follows: TBM is described as primary (congenital) if there is intrinsic collapse of the trachea and secondary (acquired) if there is extrinsic compression by the esophagus, a major vessel, or a tumor, or if the flaccid trachea resulted from infection or prolonged mechanical ventilation.[2,8] These conditions may occur together in many combinations and do not separate the causes or the treatments. For example, the primary types all have cartilage malformation as a common cause. The cartilages may be misshapen with a U or bow shape with a wide posterior membrane instead of the usual C shape, the cartilages may be maldeveloped and be very soft and collapsible, or the cartilages may just be malformed so they form a narrow plate without airway support. In this classification system, secondary types of TBM are associated with congenital anomalies, such as vascular compression from the innominate artery or aorta slightly displaced from normal positions, from vascular rings, or by the esophagus, which is often dilated in utero as in esophageal atresia (EA), resulting in compression and deformation of the trachea during development. This category also includes expanding tumors that compress the trachea, tracheal inflammation that injures and weakens the trachea, and chronic ventilation that dilates and weakens the walls of the trachea. The multiple causes and locations of TBM highlights the need for a better classification system for tracheobronchial structural integrity.

There is an association between TBM and prematurity, although the incidence has not been well studied. The incidence of bronchopulmonary dysplasia in patients with TBM is estimated to be between 50% and 72%.[1,9] Comorbid bronchopulmonary dysplasia requiring intubation at or shortly after birth may mask or delay the diagnosis of TBM. Historically, premature infants with TBM diagnosed after prolonged intubation, mechanical ventilation, and high-dose corticosteroid administration were classified as having secondary TBM. However, as intubation often occurs before a thorough airway examination, the diagnosis of primary vs secondary TBM is difficult to make. Thus, the association between TBM, prematurity, and bronchopulmonary dysplasia is recognized but remains an important area for study.

Nevertheless, primary TBM is the most common congenital anomaly of the trachea.[10] There is an association with syndromes and other congenital anomalies.[2,9] The most severe forms of TBM can result in complete collapse of the large airways and may be fatal.[2,5,10] Diagnosis is made through careful history (eg, barky cough; recurrent respiratory tract infections; BRUEs [brief, resolved, unexplained events, previously known as ALTEs [apparent life-threatening events]) with forced exhalation, laughing, and coughing; exercise intolerance; and failure to thrive due to eating and feeding disturbances) and physical examination (eg, barky cough, expiratory rhonchi); dynamic airway computed tomography (CT), which underestimates the degree of airway collapse; and dynamic, 3-phase tracheobronchoscopy, which is the standard for diagnosis of airway structure and dynamic function.[2,5,11] Medical management includes nebulizer treatments to optimize airway clearance (normal saline or hypertonic saline to thin the secretions and ipratropium bromide to decrease the secretions), minimal use of inhaled corticosteroids to avoid worsening of the airway malacia, gastroesophageal reflux disease therapy to minimize aspiration of inflammatory gastric contents, and chest physiotherapy to help with airway clearance.[2,5]

Traditional surgical management has consisted of tracheotomy to bypass or stent the affected region of the trachea, continuous positive airway pressure (CPAP) to increase the intraluminal airway pressure and improve airway distention, internal airway stents, external airway splints, and anterior aortopexy.[2,11-13] With this review, we present a novel classification scheme to facilitate discussion of patients among clinicians and describe innovative surgical techniques of anterior and posterior tracheopexy and anterior and posterior aortopexy. For the purposes of this review, we focus on pediatric TBM.

Clinical Presentation and Pathogenesis

The reported incidence of TBM is between 1 in 1445 to 1 in 2100 live births, but it has been hypothesized that this incidence may be artificially low owing to failure to make the correct diagnosis in the more severe cases and self-limited course of mild TBM.[10,14] A male predominance has been reported in 58% to 82% of cases.[5,10] Tracheobronchomalacia can present in healthy term infants but is more common in premature infants.[9]

Tracheobronchomalacia can occur in isolation, but it often presents with other congenital airway anomalies, such as laryngomalacia and laryngeal cleft, cardiopulmonary anomalies, or syndromes.[2,9] Associated syndromes can be found in the **Box**. The most common entity associated with TBM is TEF/EA.[1,6,15] Study of the relationship between TEF/EA and TM led to a proposed pathogenesis of TBM, which stems from improper foregut division into the trachea and esophagus during embryogenesis. In addition to improper midline fusion resulting in TEF and laryngeal clefts, the development of the posterior trachea may develop abnormally, ultimately resulting in the abnormal cartilage shape and wide pars membranacea often found in primary TBM.[1,6,14] An additional theory relating to TEF/EA was proposed by Davies and Cywes,[16] who suggested that dilation of the proximal esophageal pouch in utero alters the normal development of the trachea, leading to abnormal cartilage shape and a flaccid posterior trachea.

Box. Disease States Associated With Primary and Secondary Tracheobronchomalacia[a]

Primary/Congenital TM/TBM

Idiopathic TM; healthy infant

Prematurity

Cartilage congenital abnormalities
- Dyschondroplasia, chondromalacia, chondrodysplasia
- Polychondritis
- Ehlers-Danlos syndrome

Congenital syndromes
- Mucopolysaccharidosis (Hunter and Hurler syndromes)
- CHARGE syndrome/VACTERL association
- Trisomy 9 and 21
- Antley-Bixler
- Hallerman-Streiff
- Crouzon
- Pfeiffer
- Blackfan-Diamond
- Williams-Campbell
- DiGeorge
- Larsen and Larsen-like
- Brachmann de Lange
- Robin sequence
- Atelosteogenesis type 1
- Deletion 11p13, 22q11, and 12q
- Translocation 18-22
- Kneist dysplasia
- Camptomelic dysplasia

Congenital anomalies
- Tracheoesophageal fistula
- Esophageal atresia with or without laryngeal cleft
- Bronchopulmonary dysplasia

Secondary/Acquired TM/TBM

Prolonged intubation

Severe tracheobronchitis

Resulting from compression
- Vascular
  - Double aortic arch
  - Abnormal take off of the innominate artery
  - Enlarged pulmonary veins
  - Cardiac
  - Left atrial hypertrophy
  - Enlarged left atrium
  - Skeletal
  - Scoliosis
  - Pectus excavatum
  - Tumors, cysts, and masses
  - Teratomas
  - Bronchogenic cysts
  - Enterogenic cysts
  - Thymomas
  - Lymphatic malformation
  - Lymphoma
  - Neuroblastoma
  - Thyroid goiter
  - Hemangiomas

Infection
- Abscess

Posttraumatic

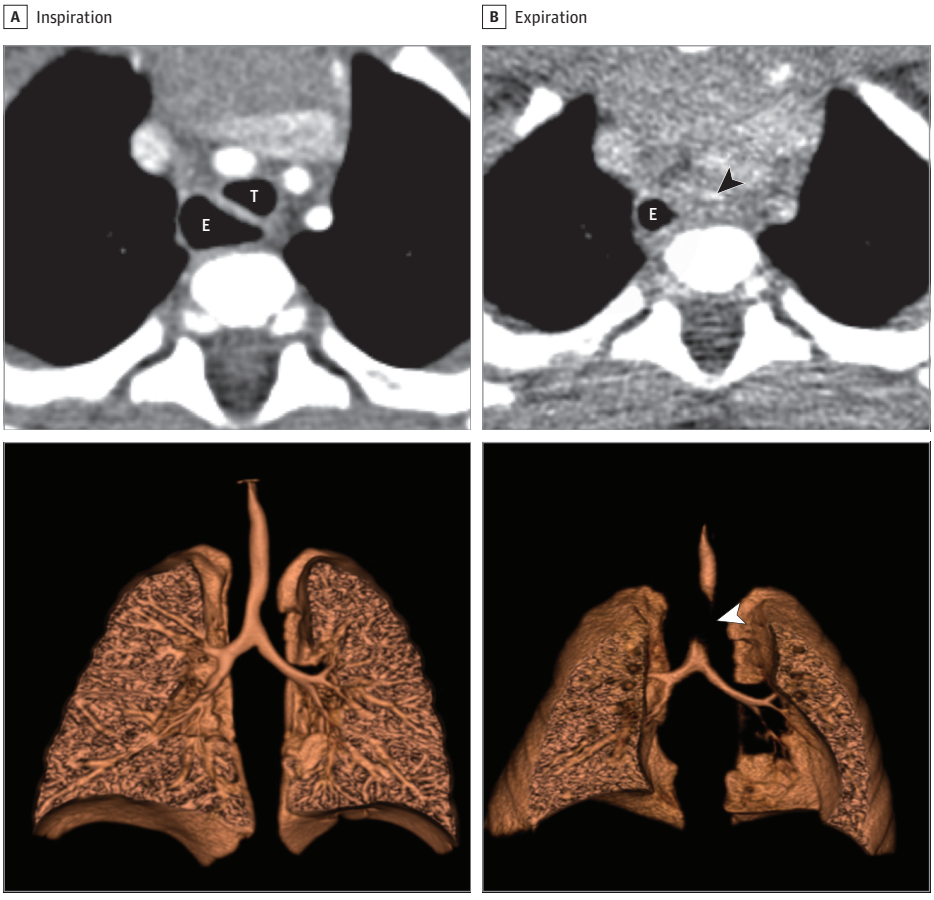
Abbreviations: CHARGE, coloboma, heart defects, choanal atresia, growth retardation, genital and/or urinary tract abnormalities, and ear anomalies; TM/TBM, tracheomalacia/tracheobronchomalacia; VACTERL, vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb anomalies.

[a] Adapted from Carden et al.[1]

Classic TBM was described as having a delayed presentation in the first 2 to 3 months of life.[2] However, other work has shown that 95% of patients with TBM have symptoms at birth.[1,14] The most common presentation includes barking cough and biphasic or expiratory stridor. Recurrent respiratory tract infections (from poor airway clearance), feeding difficulties (from esophageal dilation causing tracheal compression), spontaneous neck extension (tracheal tug opens the airway), increased work of breathing, wheezing (from small airway inflammation, which must be differentiated from reactive small airway disease), cyanosis, and apneic or dying spells (currently called ALTEs and BRUEs) may also be present.[2,5] Severe presentations of TBM may be detected at birth and require intubation and CPAP. Symptoms may be exacerbated by agitation, coughing, and feeding.[14] Tracheobronchomalacia may be diagnosed prior to TEF/EA repair by dynamic bronchoscopy, although it is often diagnosed when there is a failure to wean from CPAP and inability to extubate following esophageal atresia repair. Tracheomalacia, TBM, bronchomalacia, and small airway malacia may present as synchronous airway lesions during diagnostic and therapeutic airway endoscopies for other airway symptoms.

Tracheobronchomalacia has been divided into symptomatically mild, moderate, and severe forms. Mild forms can present with barking cough and recurrent or prolonged respiratory tract infections. This form may go undiagnosed as there is a self-limited course and the condition may resolve as the child grows, usually by age 2 years.[4,9] Improvement or resolution of symptoms may be related to the increase of airway diameter with growth of the child, allowing more air movement despite similar airway dynamic and static narrowing, thus making signs and symptoms less severe. This population is not well studied and thus is poorly understood. Moderate TBM also presents with cough and prolonged respiratory tract infections, but these patients require treatment with nebulizers and antibiotics. They miss school activities and may have exercise intolerance with easy fatigability. Patients with severe TBM present with stridor, cough, failure to thrive, recurrent pneumonias, and apneic episodes.[8] They have frequent emergency department visits and

Figure 1. Tracheal Collapse Demonstrated on Multidetector Computed Tomography



A, Esophagus (E) and trachea (T) shown on inspiration. B, Collapse of trachea during expiration, indicated with arrowheads.

hospitalizations, need for intubation, and development of bronchiectasis from recurrent and persistent lung infections. The mortality of severe TBM may be as high as 80%.[1] Tracheobronchomalacia has also been implicated as having a possible association with ALTE (now called BRUE).[17,18]

Diagnosis

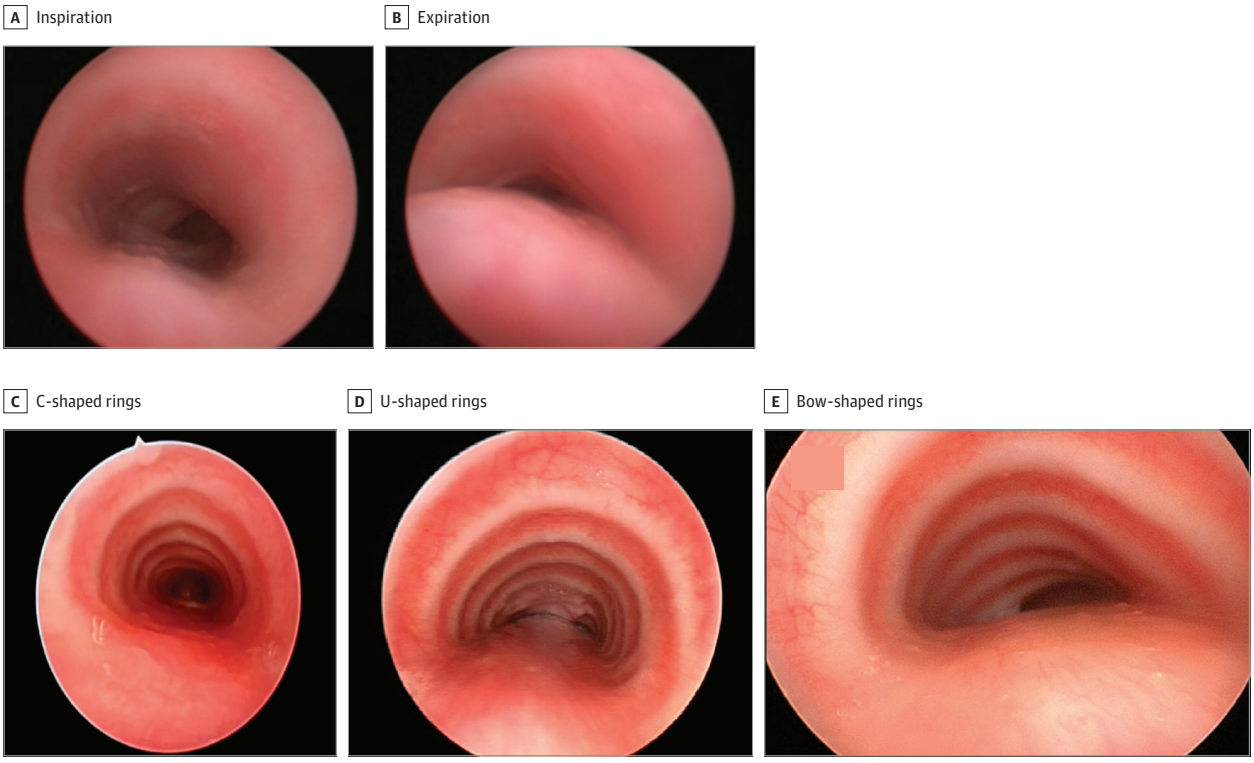
History and physical examination are paramount to suspecting the diagnosis of TBM and symptomatically classifying the severity. Expiratory stridor, barking cough, recurrent respiratory tract infections, apneic episodes, need for CPAP, and comorbid anomalies or syndromes should raise suspicion for TBM. Pulmonary function tests, plain radiographs (including inspiratory and expiratory films), tracheograms, bronchograms, airway fluoroscopy, multidetector computed tomography (MDCT), dynamic magnetic resonance imaging, and direct tracheobronchoscopy have all been used to diagnose TBM.[19-22]

At Boston Children's Hospital, we find direct tracheobronchoscopy to be of the greatest diagnostic utility. Dynamic airway MDCT can be useful, especially in evaluating collapse of the more distal airways, specifically the bronchial airways, but it underestimates the severity of the airway collapse and so cannot be used to rule out TBM. We will focus on these 2 modalities for the purposes of this review.

Multidetector computed tomography with contrast adds essential information to direct, 3-phase dynamic tracheobronchoscopy in the evaluation of TBM.[2,23,24] Multidetector computed tomography allows for traditional axial, coronal, and sagittal views with the addition of 2-dimensional and 3-dimensional (3-D) reconstructions of the thoracic structures (**Figure 1**). End-inspiration and end-expiration images can be obtained to compare airway patency. The location, characteristics, and severity of the airway collapse and airway compression can be assessed, although it is often less apparent than on dynamic bronchoscopy. Added benefits include the potential diagnosis of concomitant anomalies, such as tracheal diverticulum, vascular anomalies, pulmonary disease, and identification of the artery of Adamkiewicz, which must be preserved during thoracic surgery. If extrinsic compression is suspected or identified, MDCT is often vital for surgical planning.

In cooperative patients older than 5 years with a stable airway, these images can be obtained with the patient awake. In infants and patients with unstable airways, dynamic MDCT often requires intubation or laryngeal mask airway placement. End-inspiratory and end-expiratory phases of MDCT scanning are obtained by applying positive pressure ventilation (20 cm H_2O) to simulate breath holding during full inspiration, and by withholding positive pressure (0 cm H_2O) during expiration. Endotracheal intubation can confound results of the study by iatrogenically maintaining airway patency; thus, laryngeal mask airway or spontaneous ventilation are preferred. Ngerncham et al[23] reported the diagnostic accuracy of MDCT at 91% compared with the standard of direct tracheobronchoscopy. It is

Figure 2. Dynamic Bronchoscopic Examination of an Airway



Examination on inspiration (A) and expiration (B) exhibiting posterior wall dynamic motion resulting in near-complete obstruction of the tracheal lumen during inspiration (severe tracheomalacia). Tracheal cartilage sections shown as normal C-shaped rings (C), U-shaped rings with a wide pars membranacea demonstrating posterior intrusion (D), and bow-shaped rings, also with wide pars membranacea and posterior intrusion (E).

thought that MDCT may underestimate dynamic tracheal collapse because tracheal transmural pressure during these studies is never positive, which occurs when patients actively exhale or cough. Multidetector computed tomography was originally proposed to avoid general anesthesia, and although the procedure is brief, these patients often require sedation during MDCT, which tends to further minimize the degree of airway collapse.

Direct visualization of the airway remains the standard for diagnosing TBM.[1,2] This approach allows for the assessment of synchronous airway anomalies, such as vocal fold immobility, laryngomalacia, laryngeal cleft, laryngotracheoesophageal clefts, and TEF. The risks include those standard for direct laryngotracheobronchoscopy, including general anesthesia, and need for further studies, including MDCT, to completely characterize the TBM and understand concomitant vascular, esophageal, and thoracic anatomy as they pertain to the airway and the potential for reconstruction to open the airways.

A multidisciplinary team approach to these patients is advocated as these patients are often complex and require care coordination. A flexible fiberoptic laryngoscopy should be performed preoperatively on each patient to assess vocal fold mobility and detect laryngomalacia; this is done in clinic if possible or before induction of anesthesia in the operating room if clinic coordination is not feasible. Flexible tracheobronchoscopy performed in isolation by pediatric pulmonologists may serve to diagnose severe TBM or raise suspicion of mild to moderate TBM. Flexible tracheobronchoscopy may prompt multidisciplinary evaluation for further evaluation and management. Advantages of flexible bronchoscopy include evaluation of the distal, small airways as well as the ability to perform bronchoalveolar lavage. Rigid bronchoscopes facilitate the laryngeal examination and probing of the TEF and diverticula. As the optics and instrumentation for both modalities improve, the advantages of one technique over another diminishes in skilled hands.

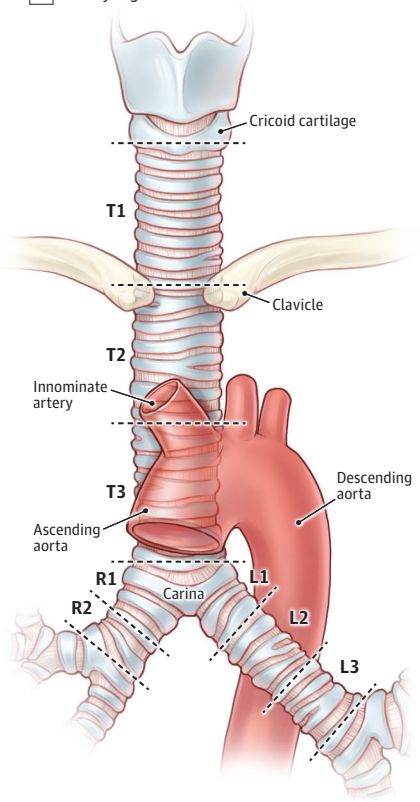
Careful preprocedural discussion with the anesthesia service is critical, as the patient must be spontaneously breathing for accurate diagnosis of TBM. Paralysis, heavy sedation, and positive pressure ventilation can mask dynamic airway collapse. A minimal amount of lidocaine hydrochloride, 1% (typically 0.5 mL), is topically applied to the true vocal folds to avoid anesthetizing the trachea, and the distal airways are examined using a Hopkins II zero-degree telescope and rigid ventilating bronchoscope of appropriate size. A dynamic, 3-phase rigid bronchoscopy is performed to completely assess the structure and function of the visible airways. The first phase of the rigid bronchoscopy is performed with the patient taking spontaneous, shallow breaths, allowing for evaluation of static compression of the trachea and bronchi. In the second phase of bronchoscopy, the aim is to see the patient vigorously breathing or coughing to assess maximum dynamic motion and collapse during expiration (**Figure 2**A and B). Achieving this activity may require tickling the airway with a fine wire or small 3F ureteral catheter (one can do this since minimal lidocaine was used to anesthetize the vocal cords and none dripped down the trachea). In the final phase of bronchos-

Figure 3. Form for Documentation of Bronchoscopy and Anatomic Divisions of the Large Airways to Facilitate Classification and Description of Tracheobronchomalacia

A Airway evaluation sheet

	FOE awake	Shallow breathing	Cough valsalva	Sedated distended pressure?	Contrast study
Tonsils					
Epiglottis		NL			
AE folds		NL			
Arytenoids		NL			
Cords		POST SCAR			
L cleft		REPAIRED			
Subglottis		10% ELLIP			
T1		"U" 10% POST	30%	FLAT	
T2		"BOW" 10% R	20%	FLAT	
T3		"BOW" 10% L	10%	FLAT	
Carina		"BOW"		FLAT	
L1		"U" 40% R	60% R	40% R	
L2		"U"	30% POST		
L3		"U"	10% POST		
R1		"U"	10%		
R2		"U"	10%		
BI		"U"	10%		
Tracheal diverticulum?		POSTERIOR CARINA		5mm DEEP	NO
TEF?					NO
Posterior artery?		NO		NO	
Secretions		THICK			
Other					
Notes			SCABBARD LMB		

B Airway segment classification



A, Standardized form for intraoperative documentation of 3-phase bronchoscopy findings, completed to correspond with **Video 1**. B, Anatomic divisions of the large airways. AE indicates aryepiglottic; BI, bronchus intermedius; ELLIP, elliptical cricoid; FOE, fiberoptic examination; L, left-sided compression; L cleft, laryngeal cleft; L1, left mainstem bronchus level 1; L2, left mainstem bronchus level 2; L3, left mainstem bronchus level 3; LMB, left mainstem bronchus. NL, normal; R, right-sided and compression; R1, right mainstem bronchus level 1; R2, right mainstem bronchus level 2; T1, trachea level 1; T2, trachea level 2; T3, trachea level 3; and TEF, tracheoesophageal fistula. U and BOW denote shape of cartilaginous rings.

copy, all secretions are removed and the airway is examined while the patient is sedated and the airway distended to 40 cm H_2O (30 cm H_2O in neonates) to dilate the trachea. The airway is carefully evaluated for structure, anatomy, and the posterior membrane is evaluated for TEF and tracheal diverticulum. Any suspicious regions are probed with an end-hole catheter and infused with water-soluble contrast to see if there is a communication with another structure, typically a TEF to the esophagus; however, other communications can occur to, for example, cysts and abscesses. The posterior glottis is probed for laryngeal cleft.

The airway is methodically evaluated by anatomic region, type of collapse, and severity of collapse. The location of the collapse—anterior, posterior, or both—is documented. The shape of cartilage is noted and is described as C (normal configuration), U, or bow shaped (Figure 2C-E). Care must always be taken to evaluate the smaller airways for malacia, either using rigid or flexible bronchoscopes. There are no standard criteria for diagnosing or describing TBM, but greater than 50% narrowing of the tracheal lumen is generally considered to be pathologic and needs to be correlated with the patient's symptoms. Most patients with symptomatic TBM will have greater than 75% narrowing on forced exhalation, and it is thought that 33% of patients with TBM will experience intermittent, complete collapse of their tracheal lumen.[7] The results of each bronchoscopy are recorded intraoperatively and dictated into the patient's record (**Figure 3**A). A sample video of a 3-phase, dynamic bronchoscopy is presented in **Video 1**.

Classification

Historically, TBM has been classified as generalized or localized disease, with studies reporting that some children had short segments of malacia. Developmental classification has since evolved into describing TBM as primary or congenital vs secondary or acquired.[1,2]

Congenital TM encompasses airway obstruction associated with flaccid cartilages and dynamic posterior membranous wall intrusion. Truly soft cartilage is rare and can be associated with immaturity or prematurity. Disease processes, such as chondrodysplasias and Ehlers-Danlos syndrome, can be associated with an intrinsic weakening of the cartilages of the tracheal wall.[1,2] Congenital TM can also be found in otherwise healthy infants and can relate to the cartilage anatomy of the trachea. If the tracheal cartilages are misshapen and do not take the common C shape, resulting in a wide pars membranacea, a greater proportion of the circumference of the trachea will be prone to dynamic collapse. Not all children with a wide pars membranacea will display clinical or bronchoscopic TBM. Tracheobronchomalacia is often seen in patients with TEF/EA, with reported rates of severe TBM in patients with EA of 11% to 33%.[2,25] Children with TEF/EA have been found to have elliptical deformities of the tracheal lumen, deficiency of the cartilaginous rings, and an increase in the width of the pars membranacea.[6]

Developmentally acquired TBM is more common than the congenital form and encompasses a variety of causes. Tracheobronchomalacia can result from severe tracheobronchitis, prolonged intubation, or trauma.[1,2] The trachea and bronchi can be extrinsically

compressed by structures such as vascular structures, thyroid goiters, congenital tumors and cysts, and the spine or sternum. Aberrant vasculature also associated with classic tracheal compression and acquired TBM. A double aortic arch results in anterior and posterior compression of the distal trachea, while a pulmonary artery sling is associated with distal anterior compression of the trachea and proximal right mainstem bronchus. Innominate artery compression typically occurs in the anterior midtrachea. Aberrant right subclavian artery is associated with posterior tracheal compression as it passes posterior to the trachea and the esophagus.[26]

We developed a classification scheme to facilitate describing individual patient variation, discussion among clinicians, and treatment planning. Evaluation with 3-phase dynamic bronchoscopy is performed as described previously and the and the findings are documented as described below.

The trachea is divided into 3 sections from proximal to distal to describe the location of malacia (Figure 3B): T1 is cricoid to the thoracic inlet, T2 is the inlet to the innominate artery, and T3 is the innominate to the top of the carina. The carina itself is evaluated independently. The left mainstem bronchi is divided into 3 sections: L1 to L3 from proximal to distal, with L2 defined as the segment where the bronchus crosses over the descending aorta. The right mainstem bronchus is divided roughly in half lengthwise, proximal to distal, and designated R1 and R2. Common structures causing compression of anatomic segments are as follows: T1 is most frequently compressed by thyroid goiters or congenital neck cysts or masses, T2 can be compressed by the innominate artery on its left anterior surface, T3 compression is often associated with the aorta anterolaterally, L1 compression is associated with the main pulmonary artery, L2 can be compressed anteriorly by the left pulmonary artery and posteriorly by the descending aorta, L3 compression is typically anterior and associated with the left pulmonary artery, and R1 and R2 are compressed anteriorly by the right pulmonary artery.

Description of the static and dynamic motion of the tracheal wall is recorded. Typically we describe the anterior and lateral percentage intrusion into the tracheal lumen and the posterior membranous wall intrusion and note any resulting coaptation. A usual notation would be T2 region pulsatile anterior collapse of 30% by innominate artery and static posterior intrusion of 30%, increasing to coaptation with coughing. Aberrant examination findings, such as aberrant right upper lobe bronchus, TEF, tracheal diverticulum, mucosal wall inflammation, friability, cobblestoning, airway masses or cysts, and airway secretions are detailed.

Treatment

At Boston Children's Hospital, patients with TBM are cared for by a multidisciplinary team, including pediatric surgeons, otolaryngologists, pulmonologists, gastroenterologists, and critical care medicine. Mild presentations of TBM can be managed without surgical intervention. Using nebulizers to optimize secretion thinning and airway clearance, and symptomatic treatment of recurrent respiratory tract infections with antibiotics and chest physiotherapy may be useful until the tracheal lumen enlarges enough to allow adequate airflow on forced expiration.[1,2,4,27,28] The malformed cartilages and the wide posterior membrane probably do not improve with age, and in fact may worsen with time and growth. These patients need to be followed up closely for progression of symptoms and to optimize treatment to avoid development of chronic lung and airway disease.

Indications for and evidence supporting the medical management of TBM are lacking. A Cochrane review published in 2012 (Goyal et al[29]) identified only a single randomized clinical trial of therapies related to the treatment of primary TBM. The study compared nebulized recombinant human deoxyribonuclease with placebo in 40 children with primary TBM and a respiratory tract infection; recombinant human deoxyribonuclease did not resolve cough at 2 weeks and subjective daytime and nighttime cough scores may have favored placebo. There were no other studies that met inclusion criteria.

Initially, a medical treatment approach to mild to moderate TBM is favored. Adjunctive therapies include hypertonic saline nebulizer treatments to thin airway secretions and facilitate clearance, inhaled low-dose corticosteroids to decrease airway inflammation and secretions, inhaled ipratropium bromide to decrease secretions, and chest physiotherapy to mobilize the secretions. Close follow-up is needed to assess response to treatment and document patients whose condition progresses symptomatically despite medical treatment.

Continuous positive airway pressure and bilevel positive airway pressure are noninvasive treatments for TBM. Both treatments increase the intraluminal pressure, and the elevated transmural pressure differential may help to open the airways, which may maintain some minimal effective tracheal patency during exhalation and cough.[1,30] However, if patients require respiratory support throughout the day, these treatments can limit oral feeding, speech development, facial development, and normal child behavior.[9,31] Historically, tracheotomy and mechanical ventilation were the mainstays of treatment for TBM. While tracheotomy facilitates oral feeding, the need for mechanical ventilation can also prevent language acquisition. In addition, pediatric tracheotomy is associated with risks and is a burden to caregivers. Tracheotomy tubes have also been used without mechanical ventilation to stent the airway and bypass the area of collapse. As would be expected, this approach results in need for custom tracheotomy tubes, frequent tube changes, and tracheal granulation tissue.[1,32]

The primary indications for surgical intervention include cyanotic and/or apneic episodes, recurrent pneumonia (>3 episodes per year), and the inability to extubate the patient owing to CPAP requirement.[2,18] Developing bronchiectasis and exercise intolerance are emerging surgical indications.[2] A single BRUE event with evidence of TBM is also an indication for hospitalization until urgent surgical repair can take place.[1,2,28]

Surgical management of TBM must consider the location, character, and degree of collapse. It is also important to consider the timing of management of comorbid conditions, particularly TEF/EA and gastroesophageal reflux disease. Extrinsic compression by surrounding structures, such as the esophagus, aberrant vasculature, or thoracic masses, must also be addressed separately or concurrently.[33,34] Occasionally, relieving vascular compression is sufficient to improve respiratory status and further interventions are not needed. However, for many patients, intrinsic collapse persists after vascular repair, necessitating additional procedures to address the primary TBM.[34]

Short-segment TM has been treated with tracheal resection and end-to-end anastomosis or slide tracheoplasty.[1,2,35,36] These strategies are not widely used because TBM rarely presents as a short-segment process amenable to this approach.[2] External tracheal splinting with autologous rib and synthetic materials has also been

documented in animal models and limited human studies.[1,2] Autologous rib has been fixed on either side of a normal cartilaginous ring to stabilize long-segment TBM.[37,38] Synthetic materials, such as Silastic meshes and ceramic rings, have also been reported, but concern for foreign body reaction, infection, and erosion have limited their use.[39,40] A newer treatment with external, custom 3-D printed splints have been reported and are currently under investigation.[13,41]

Endoscopic placement of internal tracheal stents has been attempted by multiple groups using a variety of materials, including absorbable, bare metal, and silicone. The procedure is appealing because it is less invasive and yields quicker recovery times. Unfortunately, at a minimum, stents cannot grow with the child and require future removal, dilation, or additional stent placement. Increasingly morbid complications of tracheal stents have included granulation tissue formation leading to recurrent luminal obstruction, erosion, migration, fracture, and even death.[32,42-44] At present, intraluminal stents are used only when open or endoscopic procedures are not feasible. In 2015, Morrison et al[13] described placement of an extraluminal tracheal splint using polycaprolactone, a biocompatible and bioresorbable polyester that remains in vivo for 2 to 3 years. It was created using 3-D printing technology specific to individual patient anatomy and has thus far been reported to be used only to splint severe left bronchomalacia. The splint has resulted in resolution of life-threatening airway collapse. Long-term follow-up is required, as risks of migration, erosion, foreign body reaction, and infection are still a concern, but the results have been promising.[12,13]

Until recently, anterior aortopexy was the mainstay of surgical management of intrathoracic TBM.[1,2,8,28,44] Within the deep neck and mediastinum, the trachea and mainstem bronchi are loosely attached anteriorly to the major vessels by areolar tissue and posteriorly to the mediastinal structures by connective tissue and segmental tracheal vessels. During anterior aortopexy, the thymus is often removed for exposure. The aorta and, frequently, the innominate artery, pulmonary arteries, and the pericardium are sutured to the posterior surface of the sternum (**Figure 4**A and B). This positioning creates more space in the superior mediastinum, relieves airway compression, and often opens a compressed elliptical-shaped trachea to assume a more normal circular configuration.[45] Several approaches have been described for aortopexy, partial sternotomy, thoracoscopy, and anterior and lateral thoracotomy.[46-48] Intraoperative bronchoscopy during suture placement is performed to ensure adequate decompression of the trachea at the correct locations. The placement and number of sutures is dictated by the patient's anatomy.[2] This procedure does not directly address the misshaped or flaccid cartilage of TBM or posterior membranous wall intrusion during respiration; aortopexy improves external compression of the airway by the great vessels. Anterior aortopexy has been found to improve clinical symptoms of TBM in 80% of patients but is associated with a mortality rate of 6% and a complication rate of 16.6%. Common complications included pericardial effusion, phrenic nerve injury, chylothorax, and recurrent laryngeal nerve injury.[2,49] Despite the popularity of aortopexy for TBM, there is a paucity of high-quality evidence supporting its efficacy. The literature consists mainly of small, single-center case series.[2,46-49]

Posterior aortopexy has also been described as surgical management for left mainstem bronchus compression and can be performed in conjunction with the other procedures described in this review.[46,50,51] Left mainstem bronchus collapse is most commonly associated with vascular compression. Anterior compression is attributed to the left pulmonary artery and posterior compression is associated with the descending thoracic aorta. Posterior aortopexy tacks the descending thoracic aorta to the periosteum of the thoracic vertebra. This procedure is typically performed via a right thoracotomy when there is a left descending aorta, although a left thoracotomy can be more effective. Monitoring of the lower extremity blood pressure is important to ensure there is not restriction to lower body blood flow with this maneuver.[46,50]

Anterior and posterior tracheopexy have been described as novel techniques in the surgical management of TBM.[11,52-54] Aortopexy alone was unable to address the malformed tracheal cartilages and the posterior membranous intrusion common in TBM. The procedure can be extended to tracheobronchopexy for the treatment of TBM. Posterior tracheopexy is performed by passing autologous pledgeted polypropylene sutures into but not through the posterior tracheal membrane, taking care to stay extraluminal, and securing them to the anterior longitudinal spinal ligament. Anterior tracheopexy utilizes a similar suturing technique via the anterior tracheal wall and elevates the trachea toward the posterior sternum.

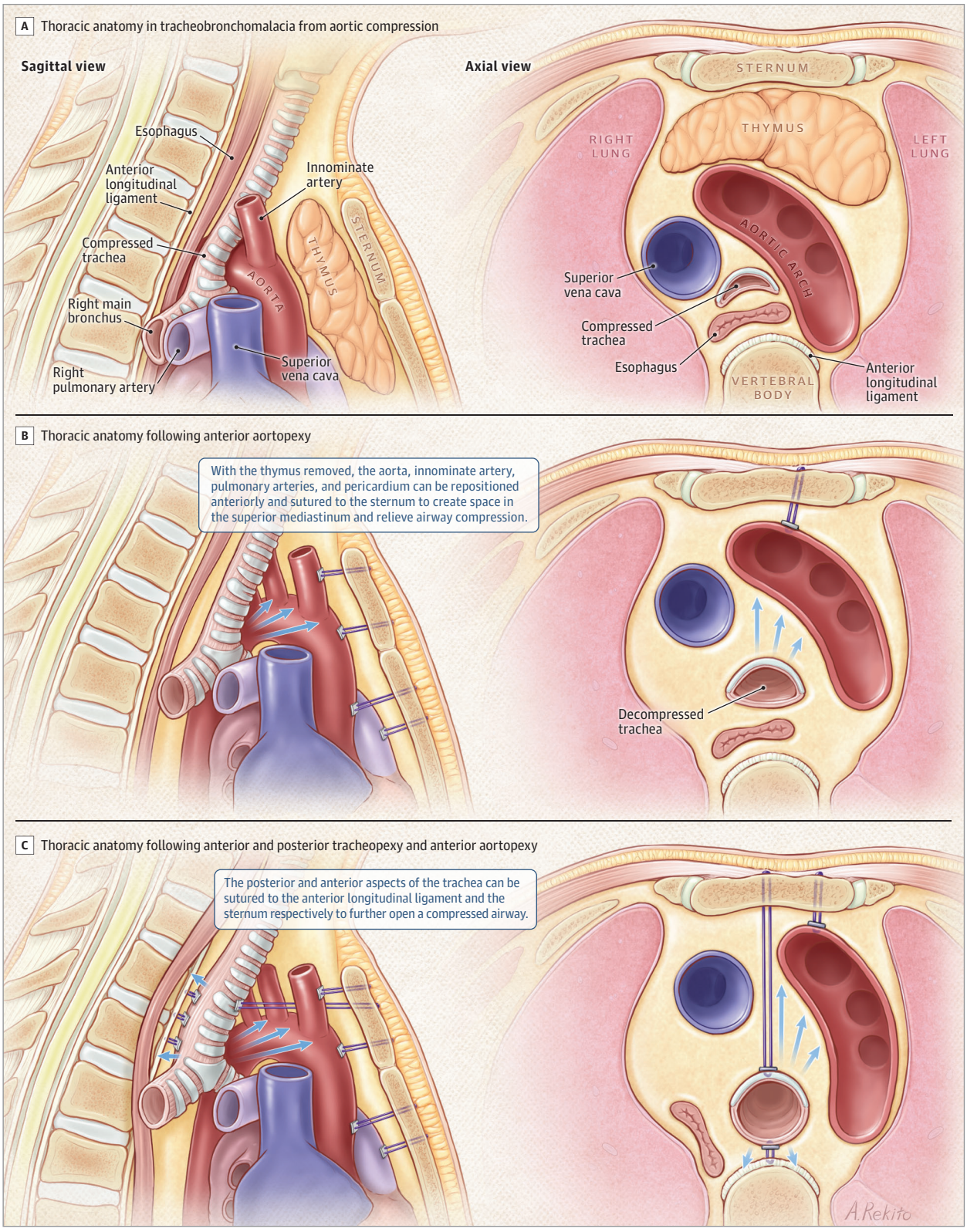
In both anterior and posterior tracheopexy, suturing is performed under direct bronchoscopic guidance to assess improvement in tracheal collapse with each suture placement and during the tying process to avoid excess tension on the trachea (Figure 4C and D; **Video 2**). Again, the number and placement of sutures are guided by the patient's anatomy. Anterior and posterior tracheopexies have been performed via median sternotomy, thoracotomy, lateral neck dissection approaches, and thoracoscopic approach, and by using a robot.[11,52-54] Aortopexy can also be performed during the same procedure. Although this technique is in its infancy and published case series consist of fewer than 100 patients from a single institution, the results have been promising. Patients have demonstrated improved exercise tolerance, less documented pneumonias, improved supplemental oxygen requirements, and resolution of their ALTE events. Complications have included bilateral vocal fold paralysis requiring tracheotomy and the need for further revision surgeries. There has been 1 death in a patient with severe congenital heart disease who developed multiorgan system failure 2 months postoperatively.[11,52] Larger studies with longer follow-up are needed to determine the true risks and benefits of these procedures.

Synchronous airway lesions, such as laryngomalacia and laryngeal cleft, are addressed at a later setting once the TBM has been treated. Ideally, an effort should be made to repeat the flexible fiberoptic laryngoscopy examination on all patients following tracheopexy to assess for vocal fold mobility. When immobility of 1 or both vocal cords is diagnosed, close follow-up by pediatric otolaryngologist is required. Immobility of one or both vocal folds that persists or is symptomatic has been managed with speech therapy, injection laryngoplasty, and tracheotomy.

Conclusions

Tracheobronchomalacia is the most common congenital tracheal anomaly and is seen with increasing frequency in tertiary care pediatric centers. Presenting with barking cough and stridor, TBM

Figure 4. Illustrations of Thoracic Structures



Thoracic structures before anterior aortopexy (A), after anterior aortopexy (B), and after anterior and posterior tracheopexy and anterior aortopexy (C). The blue arrows show the direction of pull on tracheal wall achieved by sutures being placed.

is characterized by dynamic collapse of the trachea. It is commonly associated with other airway anomalies and syndromes. Mild forms require only supportive management and may improve with age, but more severe manifestations can be associated with recurrent respiratory tract infections, chronic lung injury, bronchiectasis, need for positive-pressure ventilation, ALTE, and death. A multidisciplinary team approach to diagnosis and management of TBM is ideal. Workup includes flexible fiberoptic laryngoscopy, MDCT, and dynamic laryngotracheobronchoscopy. Classification and severity staging facilitate case discussions among clinicians and guide surgical management, when indicated. There is no consensus on the ideal surgical approach to TBM; anterior tracheopexy, posterior tracheopexy, and posterior aortopexy are the latest techniques, joining anterior aortopexy and stents, and show promise.

Pediatric otolaryngologists should have an increased awareness of TBM, as its presenting symptoms will often prompt referral to otolaryngologist. Pediatric otolaryngologists should incorporate dynamic bronchoscopy into endoscopic airway evaluation in patients with risk factors for TBM or symptoms suspicious for TBM. Pediatric otolaryngologists are critical members of the multidisciplinary teams that manage care for patients with TBM.

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REFERENCES

1. Carden KA, Boiselle PM, Waltz DA, Ernst A. Tracheomalacia and tracheobronchomalacia in children and adults: an in-depth review. *Chest*. 2005;127(3):984-1005. doi:10.1378/chest.127.3.984

2. Fraga JC, Jennings RW, Kim PCW. Pediatric tracheomalacia. *Semin Pediatr Surg*. 2016;25(3):156-164. doi:10.1053/j.sempedsurg.2016.02.008

3. Holinger PH, Johnston KC, Parchet VN, Zimmermann AA. Congenital malformations of the trachea, bronchi and lung. *Ann Otol Rhinol Laryngol*. 1952;61(4):1159-1180. doi:10.1177/000348945206100419

4. Baxter JD, Dunbar JS. Tracheomalacia. *Ann Otol Rhinol Laryngol*. 1963;72:1013-1023. doi:10.1177/000348946307200415

5. Deacon JWF, Widger J, Soma MA. Paediatric tracheomalacia—a review of clinical features and comparison of diagnostic imaging techniques. *Int J Pediatr Otorhinolaryngol*. 2017;98:75-81. doi:10.1016/j.ijporl.2017.04.027

6. Wailoo MP, Emery JL. The trachea in children with tracheo-oesophageal fistula. *Histopathology*. 1979;3(4):329-338. doi:10.1111/j.1365-2559.1979.tb03014.x

7. Wittenborg MH, Gyepes MT, Crocker D. Tracheal dynamics in infants with respiratory distress, stridor, and collapsing trachea. *Radiology*. 1967;88(4):653-662. doi:10.1148/88.4.653

8. Benjamin B, Cohen D, Glasson M. Tracheomalacia in association with congenital tracheoesophageal fistula. *Surgery*. 1976;79(5):504-508.

9. Jacobs IN, Wetmore RF, Tom LW, Handler SD, Potsic WP. Tracheobronchomalacia in children. *Arch Otolaryngol Head Neck Surg*. 1994;120(2):154-158. doi:10.1001/archotol.1994.01880260026006

10. Boogaard R, Huijsmans SH, Pijnenburg MW, Tiddens HA, de Jongste JC, Merkus PJ. Tracheomalacia and bronchomalacia in children: incidence and patient characteristics. *Chest*. 2005;128(5):3391-3397. doi:10.1378/chest.128.5.3391

11. Bairdain S, Zurakowski D, Baird CW, Jennings RW. Surgical treatment of tracheobronchomalacia: a novel approach. *Paediatr Respir Rev*. 2016;19:16-20.

12. Zopf DA, Hollister SJ, Nelson ME, Ohye RG, Green GE. Bioresorbable airway splint created with a three-dimensional printer. *N Engl J Med*. 2013;368(21):2043-2045. doi:10.1056/NEJMc1206319

13. Morrison RJ, Hollister SJ, Niedner MF, et al. Mitigation of tracheobronchomalacia with 3D-printed personalized medical devices in pediatric patients. *Sci Transl Med*. 2015;7(285):285ra64. doi:10.1126/scitranslmed.3010825

14. Mair EA, Parsons DS. Pediatric tracheobronchomalacia and major airway collapse. *Ann Otol Rhinol Laryngol*. 1992;101(4):300-309. doi:10.1177/000348949210100403

15. Blair GK, Cohen R, Filler RM. Treatment of tracheomalacia: eight years' experience. *J Pediatr Surg*. 1986;21(9):781-785. doi:10.1016/S0022-3468(86)80366-9

16. Davies MRQ, Cywes S. The flaccid trachea and tracheoesophageal congenital anomalies. *J Pediatr Surg*. 1978;13(4):363-367. doi:10.1016/S0022-3468(78)80455-2

17. Schwartz MZ, Filler RM. Tracheal compression as a cause of apnea following repair of tracheoesophageal fistula: treatment by aortopexy. *J Pediatr Surg*. 1980;15(6):842-848. doi:10.1016/S0022-3468(80)80290-9

18. Weber TR, Keller MS, Fiore A. Aortic suspension (aortopexy) for severe tracheomalacia in infants and children. *Am J Surg*. 2002;184(6):573-577. doi:10.1016/S0002-9610(02)01054-1

19. Feist JH, Johnson TH, Wilson RJ. Acquired tracheomalacia: etiology and differential diagnosis. *Chest*. 1975;68(3):340-345. doi:10.1378/chest.68.3.340

20. Lee KS, Sun MRM, Ernst A, Feller-Kopman D, Majid A, Boiselle PM. Comparison of dynamic expiratory CT with bronchoscopy for diagnosing airway malacia: a pilot study. *Chest*. 2007;131(3):758-764. doi:10.1378/chest.06-2164

21. Faust RA, Remley KB, Rimell FL. Real-time, cine magnetic resonance imaging for evaluation of the pediatric airway. *Laryngoscope*. 2001;111(12):2187-2190. doi:10.1097/00005537-200112000-00022

22. McCubbin M, Frey EE, Wagener JS, Tribby R, Smith WL. Large airway collapse in bronchopulmonary dysplasia. *J Pediatr*. 1989;114(2):304-307. doi:10.1016/S0022-3476(89)80802-9

23. Ngerncham M, Lee EY, Zurakowski D, Tracy DA, Jennings R. Tracheobronchomalacia in pediatric patients with esophageal atresia: comparison of diagnostic laryngoscopy/bronchoscopy and dynamic airway multidetector computed tomography. *J Pediatr Surg*. 2015;50(3):402-407. doi:10.1016/j.jpedsurg.2014.08.021

24. Lee EY, Boiselle PM. Tracheobronchomalacia in infants and children: multidetector CT evaluation. *Radiology*. 2009;252(1):7-22. doi:10.1148/radiol.2513081280

25. Spitz L, Kiely E, Brereton RJ. Esophageal atresia: five year experience with 148 cases. *J Pediatr Surg*. 1987;22(2):103-108. doi:10.1016/S0022-3468(87)80420-7

26. Rogers DJ, Cunnane MB, Hartnick CJ. Vascular compression of the airway: establishing a functional diagnostic algorithm. *JAMA Otolaryngol Head Neck Surg*. 2013;139(6):586-591. doi:10.1001/jamaoto.2013.3214

27. Vasko JS, Ahn C. Surgical management of secondary tracheomalacia. *Ann Thorac Surg*. 1968;6(3):269-272. doi:10.1016/S0003-4975(10)66023-7

28. Filler RM, Messineo A, Vinograd I. Severe tracheomalacia associated with esophageal atresia: results of surgical treatment. *J Pediatr Surg*. 1992;27(8):1136-1140. doi:10.1016/0022-3468(92)90575-R

29. Goyal V, Masters IB, Chang AB. Interventions for primary (intrinsic) tracheomalacia in children. *Cochrane Database Syst Rev*. 2012;10:CD005304.

30. Panitch HB, Allen JL, Alpert BE, Schidlow DV. Effects of CPAP on lung mechanics in infants with acquired tracheobronchomalacia. *Am J Respir Crit Care Med*. 1994;150(5 Pt 1):1341-1346. doi:10.1164/ajrccm.150.5.7952562

31. Wiseman NE, Duncan PG, Cameron CB. Management of tracheobronchomalacia with

continuous positive airway pressure. *J Pediatr Surg*. 1985;20(5):489-493. doi:10.1016/S0022-3468(85)80471-1

32. Fauroux B, Lavis JF, Nicot F, et al. Facial side effects during noninvasive positive pressure ventilation in children. *Intensive Care Med*. 2005;31(7):965-969. doi:10.1007/s00134-005-2669-2

33. Wright CD, Graham BB, Grillo HC, Wain JC, Mathisen DJ. Pediatric tracheal surgery. *Ann Thorac Surg*. 2002;74(2):308-313. doi:10.1016/S0003-4975(02)03613-5

34. Fraga JC, Calkoen EE, Gabra HOS, McLaren CA, Roebuck DJ, Elliott MJ. Aortopexy for persistent tracheal obstruction after double aortic arch repair. *J Pediatr Surg*. 2009;44(7):1454-1457. doi:10.1016/j.jpedsurg.2009.03.035

35. Greenholz SK, Karrer FM, Lilly JR. Contemporary surgery of tracheomalacia. *J Pediatr Surg*. 1986;21(6):511-514. doi:10.1016/S0022-3468(86)80222-6

36. Grillo HC, Zannini P. Management of obstructive tracheal disease in children. *J Pediatr Surg*. 1984;19(4):414-416. doi:10.1016/S0022-3468(84)80265-1

37. Meyer R. New concepts in laryngotracheal reconstruction. *Trans Am Acad Ophthalmol Otolaryngol*. 1972;76(3):758-765.

38. Johnston MR, Loeber N, Hillyer P, Stephenson LW, Edmunds LH Jr. External stent for repair of secondary tracheomalacia. *Ann Thorac Surg*. 1980;30(3):291-296. doi:10.1016/S0003-4975(10)61260-X

39. Rainer WG, Newby JP, Kelble DL. Long term results of tracheal support surgery for emphysema. *Dis Chest*. 1968;53(6):765-772. doi:10.1378/chest.53.6.765

40. Amedee RG, Mann WJ, Lyons GD. Tracheomalacia repair using ceramic rings. *Otolaryngol Head Neck Surg*. 1992;106(3):270-274. doi:10.1177/019459989210600313

41. Huang L, Wang L, He J, et al. Tracheal suspension by using 3-dimensional printed personalized scaffold in a patient with tracheomalacia. *J Thorac Dis*. 2016;8(11):3323-3328. doi:10.21037/jtd.2016.10.53

42. Filler RM, Forte V, Chait P. Tracheobronchial stenting for the treatment of airway obstruction. *J Pediatr Surg*. 1998;33(2):304-311. doi:10.1016/S0022-3468(98)90452-3

43. Furman RH, Backer CL, Dunham ME, Donaldson J, Mavroudis C, Holinger LD. The use of balloon-expandable metallic stents in the treatment of pediatric tracheomalacia and bronchomalacia. *Arch Otolaryngol Head Neck Surg*. 1999;125(2):203-207. doi:10.1001/archotol.125.2.203

44. de Trey LA, Dudley J, Ismail-Koch H, et al. Treatment of severe tracheobronchomalacia: ten-year experience. *Int J Pediatr Otorhinolaryngol*. 2016;83:57-62. doi:10.1016/j.ijporl.2016.01.022

45. Filler RM, Rossello PJ, Lebowitz RL. Life-threatening anoxic spells caused by tracheal compression after repair of esophageal atresia: correction by surgery. *J Pediatr Surg*. 1976;11(5):739-748. doi:10.1016/0022-3468(76)90098-1

46. Jennings RW, Hamilton TE, Smithers CJ, Ngerncham M, Feins N, Foker JE. Surgical approaches to aortopexy for severe tracheomalacia. *J Pediatr Surg*. 2014;49(1):66-70. doi:10.1016/j.jpedsurg.2013.09.036

47. Abdel-Rahman U, Ahrens P, Fieguth HG, Kitz R, Heller K, Moritz A. Surgical treatment of tracheomalacia by bronchoscopic monitored aortopexy in infants and children. *Ann Thorac Surg*. 2002;74(2):315-319. doi:10.1016/S0003-4975(02)03642-1

48. Brawn WJ, Huddart SN. Tracheoaortopexy via midline sternotomy in tracheomalacia. *J Pediatr Surg*. 1991;26(6):660-662. doi:10.1016/0022-3468(91)90004-D

49. Calkoen EE, Gabra HO, Roebuck DJ, Kiely E, Elliott MJ. Aortopexy as treatment for tracheo-bronchomalacia in children: an 18-year single-center experience. *Pediatr Crit Care Med*. 2011;12(5):545-551. doi:10.1097/PCC.0b013e3182070f6f

50. Arcieri L, Serio P, Nenna R, et al. The role of posterior aortopexy in the treatment of left mainstem bronchus compression. *Interact Cardiovasc Thorac Surg*. 2016;23(5):699-704. doi:10.1093/icvts/ivw209

51. Hungate RG, Newman B, Meza MP. Left mainstem bronchial narrowing: a vascular compression syndrome? evaluation by magnetic resonance imaging. *Pediatr Radiol*. 1998;28(7):527-532. doi:10.1007/s002470050404

52. Shieh HF, Smithers CJ, Hamilton TE, et al. Posterior tracheopexy for severe tracheomalacia. *J Pediatr Surg*. 2017;52(6):951-955. doi:10.1016/j.jpedsurg.2017.03.018

53. Bjornson C, Brindle M, Bailey JM, Mitchell I, Soles M. Delayed diagnosis of high proximal tracheoesophageal fistula in esophageal atresia and a novel approach to the treatment of tracheomalacia by submanubrial tracheopexy. *Springerplus*. 2014;3:113. doi:10.1186/2193-1801-3-113

54. Bairdain S, Smithers CJ, Hamilton TE, et al. Direct tracheobronchopexy to correct airway collapse due to severe tracheobronchomalacia: Short-term outcomes in a series of 20 patients. *J Pediatr Surg*. 2015;50(6):972-977. doi:10.1016/j.jpedsurg.2015.03.016