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Current concepts in tracheobronchomalacia: diagnosis and treatment

Ali Kamran, Benjamin Zendejas, Russell W. Jennings*

Esophageal and Airway Treatment Center, Department of Pediatric General Surgery, Boston Children's Hospital, Harvard Medical School, Boston, MA, United States

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ABSTRACT

Airway collapse from dynamic tracheobronchomalacia (TBM), static compression from vascular compression, and/or tracheobronchial deformation are challenging conditions. Patients are best assessed and managed by a multidisciplinary team in centers specializing in complex pediatric airway disorders. Suspicion is made through clinical history and physical examination, diagnosis of location and severity by dynamic 3-phase bronchoscopy, and surgical treatment planning by MDCT and other studies as necessary to completely understand the problems. The treatment plan should be patient-based with a thorough approach to the underlying pathology, clinical concerns, and combined abnormalities. Patients should undergo maximum medical therapy prior to committing to other interventions. For those children considered candidates for surgical intervention, all other associated conditions, including vascular anomalies, chest wall deformities, mediastinal lesions, or other airway pathologies, should also be considered. Our preference is to correct the airway lesions at the same operation as other comorbidities, if possible, to prevent multiple reoperations with their attendant increased risks. We also strongly advocate for the use of recurrent laryngeal nerve monitoring in all cases of cervical or thoracic surgery to minimize the risks to vocal cord function and laryngeal sensation. Studies that evaluate the effect of these interventions on the patient and caregiver's quality of life are needed to fully grasp the impact of TBM on this challenging patient population.

Introduction

Tracheobronchomalacia (TBM) is a relatively common abnormal condition of the respiratory tract characterized by an excessive dynamic collapse of the trachea and main bronchi during expiration.

The normal trachea and main bronchi are supported by relatively rigid C-shaped cartilages and a narrow pliable posterior membrane. During expiration and coughing, the posterior membrane bulges inward, resulting in dynamic airway narrowing. Thus, a certain degree of airway narrowing caused by the posterior membrane intrusion is physiological and accelerates airflow and mucus clearance [1-3]. In healthy individuals, the decrease in airway diameter caused by posterior membrane intrusion is typically 10-20%, not more than 50% even during forced expiratory maneuvers such as coughing [3,4]. In patients with TBM, airway cartilages are often developed in an aberrant inverted "U" or "bow" shape (Fig. 1), leaving the posterior membrane broader and more mobile [5]. Thus, normal changes in airway caliber during breathing are accentuated, and dynamic airway collapse occurs during forced expiration and coughing due to increased intrathoracic pressure [3].

Dynamic airway collapse (TBM) can present in isolation or in association with external airway compression (often static). Because of the close anatomical relationship between intrathoracic structures, airway compression can be caused by surrounding structures, typically the blood vessels and the esophagus. In patients with a lack of normal cartilaginous stiffness, the airway structure becomes softer and more susceptible to collapse [5-7]. Congenital weakness (or the exceedingly rare absence) of cartilage rings, prolonged positive pressure ventilation, chronic infectious/inflammatory conditions, or continued exposure to steroids can compromise airway integrity and rigidity [2,4-7]. Abnormal position or anatomy of the blood vessels (such as seen in patients with vascular rings or aberrant arteries) [8-14] or decrease in the thoracic cavity (such as seen in patients with mediastinal masses, chest wall deformities, or anterior spinal displacement) can also have profound effects on external airway compression (Fig. 2).

Clinical presentation

The clinical features are non-specific and depend on the location, extent, and severity of the airway collapse, as well as other

* Corresponding author at: Esophageal and Airway Treatment (EAT) Center, Boston Children's Hospital, 300 Longwood Ave. Boston, MA 02115, United States.
E-mail address: russell.jennings@childrens.harvard.edu (R.W. Jennings).

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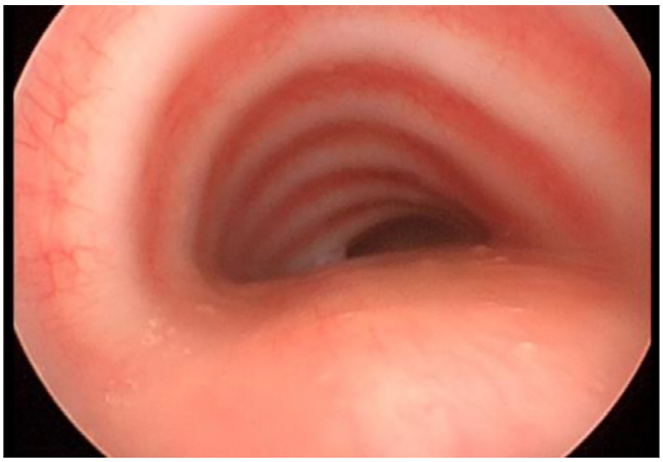

Fig. 1. Tracheal bow-shaped cartilage rings with a wide posterior membrane intruded excessively during expiration, resulting in severe airway collapse.

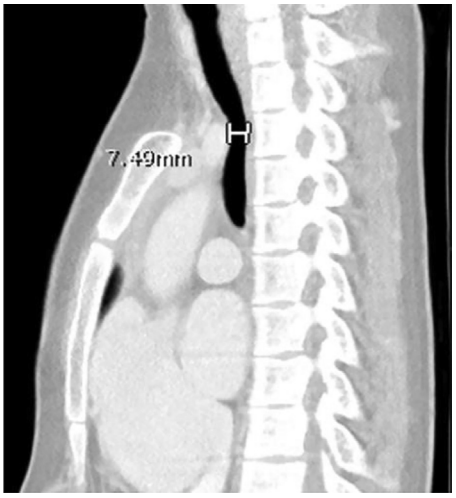

Fig. 2. Severely narrowed anterior-posterior diameter between the spine and sternum, resulting in airway compression caused by blood vessel. In such cases, the only option is to increase the space between the sternum and the spine to improve airway compression.

factors that may not yet be identified. Long-segment, multi-level (affecting major and minor airways) or severe (complete collapse) TBM is typically evident clinically from birth; however, symptoms may not appear until two or three months of age or later in some cases [2,4]. The consequences of TBM encompasses a spectrum of signs and symptoms, ranging from expiratory stridor with a typical "barking" cough (so-called "TOF cough" in Europe), rhonchorous breathing, exercise intolerance, feeding difficulties with shortness of breath or cough, prolonged/recurrent pulmonary infections, development of bronchiectasis, respiratory distress episodes and life-threatening events [2,4-7,15]. Patients with static airway collapse alone, caused by external compression and/or tracheobronchial deformation, may have similar symptoms without a barking cough, and static airway stenosis may appear with biphasic stridor.

In patients with TBM, airway closure during cough and ineffective cough due to an underlying condition can cause impaired clearance of secretion [2,4]. Hence, they are at increased risk of frequent upper respiratory illness, prolonged recovery from upper respiratory infections, more severe respiratory infections, recurrent/persistent pneumonia, and bronchiectasis [16-18]. The airway collapse can also result in insufficient air movement and ventilation; therefore, patients may experience exercise intolerance, hypoxic episodes, or apneic events–often initiated or aggravated by activities or conditions that increase the intrathoracic pressure [4].

Evaluation

Endoscopic assessment

Endoscopic evaluation of the airway is the most critical component of assessing children with a high index of suspicion for dynamic and/or static airway collapse based on the clinical history and physical examination.

Our team at the Esophageal and Airway Treatment (EAT) Center at Boston Children's Hospital has introduced 3-phase dynamic bronchoscopy (rigid or flexible) as a useful diagnostic tool for a complete assessment of the structure and function of the visible airways [5,19] (Fig. 3). The first phase is performed while the patient is taking spontaneous and shallow breathing to assess static airway compression and tracheobronchial deformation and/or lesions. The second phase is to induce coughing and Valsalva maneuvers while observing the entire airway again, to assess maximum dynamic motion and collapse during expiration (Fig. 3A-C), as well as the secretion accumulation that gets displaced from the distal airways and comes into the larger airways (secretions should be sent for culture). This dynamic phase is critical for identifying the regions and severity of TBM. Clear communication with the anesthesiologist is essential during the dynamic portion so that the desired level of sedation can be titrated to allow the patient to cough while maintaining safe control of the airway. Concerns about tracheal injury have not been realized using this technique in over 1000 dynamic bronchoscopies. The third phase is to distend the airways to 40–60 cm of water after aspirating all secretions out to reveal the anatomic abnormalities that are best seen with airway distention maneuvers, such as tracheal diverticulum (Fig. 3D), tracheoesophageal fistula, and aberrant bronchi, as well as regions of static compression or malformation.

Our team routinely uses a standardized reporting system for endoscopic evaluation based on anatomic regions and the severity of airway collapse [19-21]. Anatomic regions are classified into the upper (T1), middle (T2), and lower (T3) trachea; and right and left mainstem bronchi. T1 is the upper third of the trachea, located above the clavicles and up to the cricoid cartilage (extrathoracic trachea). T2 is the middle third of the trachea, located below the clavicles to the takeoff of the innominate artery, which can usually be easily visualized during bronchoscopy. T3 is the lower third of the trachea, the carina is the region between and the takeoff of the two mainstem bronchi. The right mainstem is divided into the proximal and distal right mainstem (R1, R2), and the left mainstem is divided into the proximal, middle, and distal left mainstem (L1, L2, L3). For each of these regions, we determine the percentage of airway narrowing and contribution of anterior/lateral compression and/or posterior or anterior intrusion, as well as note the shape of the cartilages (C, U, or bow) and any other airway distortion or lesions.

Most surgeons typically define TBM as a greater than 50% expiratory reduction in the cross-sectional luminal area [4]. However, bronchoscopic findings must be correlated with clinical features to be considered pathologic. Children with symptomatic TBM are often associated with more than 75% closure of the airway in one or more regions during forced exhalation or coughing, and in those presenting recurrent pulmonary infections and hypoxic episodes, typically, the airway is completely closed in one or more regions during forced exhalation or coughing [2,5].

A dynamic 3-phase flexible bronchoscopy with a small scope can be helpful if there is a concern for small airway collapse, which is often found in premature infants and children with bronchopulmonary dysplasia [22,23]. The collapse of the lobar or segmen-

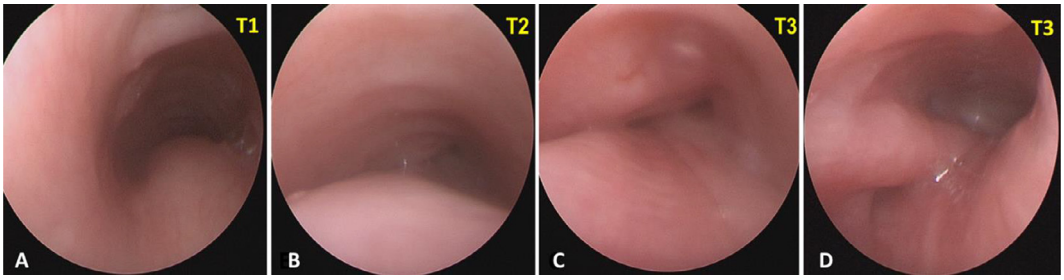

Fig. 3. Dynamic 3-phase bronchoscopy, showing multi-level airway collapse from TBM and vascular compression, and tracheal diverticulum after EA/TEF. A. Normal upper trachea with 30% posterior intrusion during phase 2 coughing; B. TBM at mid trachea with 50% closure of airway from posterior intrusion during phase 2 coughing; C: Antero-lateral 40% compression caused by right aortic arch at lower trachea with 50% posterior intrusion during phase 2 coughing; D. Tracheal diverticulum at lower trachea during phase 3 distention. (T1: upper trachea; T2: mid trachea; T3: lower trachea)

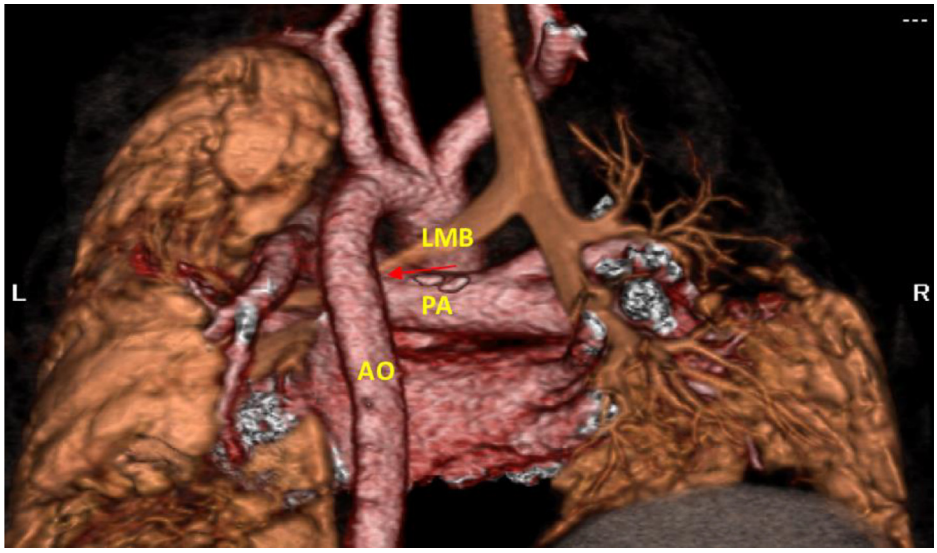

Fig. 4. Left mainstem bronchus (LMB) trapped between descending aorta (AO) and pulmonary artery (PA)

tal airways with expiration or removal of positive pressure may indicate no surgical solution and require the need for prolonged positive pressure and avoidance of steroids to keep these airways open. Occasionally, we see children with both large and small airway collapse. Those patients that have a more severe collapse of the large airways at lower positive airway pressures may benefit from surgical interventions that seek to support or open the trachea and mainstem bronchi, although this is an area of active investigation.

Multi-detector computed tomography (MDCT)

MDCT scan with three-dimensional reconstructions of the airway and surrounding vasculature is a very useful tool for preoperative evaluation [24-26]. This diagnostic utility can be helpful in identifying abnormal position or anatomy of the great vessels, as well as measuring the distance between the anterior-most part of the descending aorta and the anterior longitudinal ligament of the spine. If the descending aorta is located too far anteriorly, the mid-portion of the left mainstem bronchus (L2) can be trapped between the descending aorta and pulmonary artery, resulting in narrowing the bronchus (Fig. 4) [27]. In this case, a descending posterior aortopexy is often performed. Identification of the Artery of Adamkiewicz during the MDCT scan guides the surgeon in avoiding injury to this critical artery during a posterior descending aortopexy and mediastinal dissection. The location, characteristics, and severity of the static airway narrowing and compression (but not dynamic TBM) can be assessed using MDCT scan, although it typically underestimates the severity of the static airway collapse and cannot be used to rule out TBM.

Further studies

Patients with a history of apneic episodes should be evaluated for cardiac and neurologic causes. Other studies that can be helpful in the pre-operative assessment include an echocardiogram (to confirm proper heart function prior to the surgical intervention), ventilation-perfusion (V/Q) scan (to assess lung performance), esophagogram (to assess swallowing, strictures, aspiration), and nuclear clearance studies (available in some facilities to assess the function of the esophagus, emptying of the stomach, aspiration, and tracheal clearance).

Medical management

Clinical presentation in combination with bronchoscopic findings helps to determine indications and timing for treatment. Currently, there are no generally accepted standard strategies regarding the medical and surgical management of patients with symptomatic TBM. There is the opinion among many physicians that symptoms may resolve within a few years without surgical intervention. The airway with a lack of normal cartilaginous stiffness tends to become more rigid with continued growth and avoidance of steroids and infections, and minor degrees of airway collapse may be expected to improve by age 1-2 years with growth [3]. However, congenital cartilaginous malformation or deformation typically do not improve with age, and in fact, may worsen

with time and growth. These patients need to be followed up closely for the progression of symptoms. Patients affected by mild to severe TBM may benefit from medical management while awaiting airway structural stability and luminal enlargement. However, indications for and evidence supporting the medical treatment of TBM are lacking [28]. In our opinion, all patients need to be optimized using medical therapy prior to considering surgical correction if that is an option, at a minimum to control secretions and infections, as well as optimum nutrition and at least two weeks of adequate growth and positive nitrogen balance in order to tolerate the stress of surgery. At the EAT Center, the mainstay of medical management is the optimization of mucociliary clearance since the airway clearance mechanism is altered by airway collapse. Ipratropium bromide (Atrovent®) is administered to decrease secretions without thickening the secretions and may help to stiffen the smaller airways [29]. Normal saline or hypertonic saline is nebulized 1-3 times a day to loosen and thin the airway secretions, which are more easily transported by the ciliary clearance mechanism. Pulmonary hygiene and chest physiotherapy can also help to enhance mucociliary clearance. Low-dose inhaled corticosteroids may decrease airway mucosal swelling from inflammation and decrease airway secretions; however, routine and aggressive or continuous use of steroids should be avoided due to adverse effects on cartilage development. Continued exposure to steroids can be associated with the risks of cartilage degradation and tissue weakening; therefore, it may cause progressive tracheomalacia and/or small airway collapse [30]. Besides, the enthusiastic use of steroids can lead to Cushingoid appearance and adrenal suppression. The early use of antibiotics during an active respiratory infection may decrease the severity and length of symptoms, so they should be started early in those patients with recurrent infections. Control of gastroesophageal reflux (GER) to minimize aspiration of inflammatory gastric contents is also encouraged [31]. Patients with vascular rings and aberrant subclavian arteries tend to have esophageal compression, and this may need to be addressed as well at the time of surgery. In patients with a history of Esophageal Atresia (EA), the concerns for esophageal dysmotility with bacterial or fungal overgrowth and/or GER, as well as recurrent or congenital tracheoesophageal fistula (TEF), should also be considered. It is very important to understand the laryngeal anatomy, including laryngeal clefts, and the vocal cord function, prior to contemplating surgery.

Surgical interventions

Surgical treatment is reserved for symptomatic patients with bronchoscopic findings of severe TBM and/or airway compression who fail maximum medical management. The surgical plan in each patient must be individualized based on the type and location of airway collapse, as well as all other associated conditions if present, such as airway cartilaginous deformation, mediastinal masses, abnormal vasculature anatomy or position, tracheoesophageal fistula, abnormal airway branching, as well as chest wall and/or spine deformities.

Multiple options are available for surgical intervention, including anterior aortopexy, anterior and posterior tracheobronchopexy, posterior descending aortopexy, tracheobronchial resection and reconstruction, external splint, and intraluminal stent, as well as vascular reconstruction in some uncommon cases, and rarely chest wall expansion.

Tracheostomy

In the past, tracheostomy and long-term medical ventilation was the mainstay surgical approach in patients with severe TBM. The utility of the tracheostomy is to provide a mechanism for the delivery of positive airway pressure, secretion suctioning and removal, and a degree of airway stenting. However, it is not a risk-free procedure, particularly in small infants, requiring changes in the size and length of the tracheostomy tube as the child grows. Placement of tracheostomy is associated with considerable risks, including tracheal injury, inflammation-causing granulation tissue, tracheo-arterial fistula formation, tracheal stenosis, tracheal pouch formation, tracheo-esophageal fistula formation, tracheal plugging, accidental decannulation, delayed vocalization, and difficulty with decannulation as it does not address the problem of the collapsible airway distal to the end of the tracheostomy tube. Furthermore, secondary TBM and tracheal fibrosis from the presence of the tracheostomy are not uncommon. Thus, this approach should be avoided if possible and reserved as a last resort in highly selected patients with life-threatening airway obstruction who have failed other therapeutical strategies or in whom the risk-benefit analysis points to tracheostomy placement.

Aortopexy

Until recently, aortopexy has been the mainstay of surgical treatment for intrathoracic TBM and still a favored option in many centers [32-38]. During this procedure, the thymus is often removed or displaced to create space in the superior mediastinum and to relieve anterior pressure on the trachea, the ascending aorta or arch (and frequently innominate artery, pulmonary arteries, and/or pericardium) is pulled anteriorly and sutured to the posterior surface of the sternum. The large airways are loosely attached anteriorly to the major blood vessels by areolar tissue. Aortopexy supports the anterior wall of the airway when the areolar attachments of the aorta and innominate artery remain intact. The areolar tissue may grow and relax with time, and therefore the structures collapse back down, or the aortopexy sutures may “cheese wire” through the aorta, and the vessels may fall back. While aortopexy may relieve anterior airway compression from the great vessels, it does not directly address the flaccid or malformed cartilages nor dynamic airway collapse caused by the posterior intrusion, which is the major contributor to airway collapse in many pediatric cases [39].

Tracheobronchopexy (anterior, posterior)

Our team at the EAT Center has described the novel techniques of posterior and anterior tracheobronchopexy to directly address the airway collapse from posterior intrusion and anterior compression, respectively [16,20-22,33]. Posterior tracheobronchopexy is the most common first procedure for posterior intrusion type TBM. A pre-op chest x-ray or MDCT scan can reveal the optimal interspace and side for entry to expose the critical portions of airway collapse. Usually, the 3rd or 4th interspace is chosen on the side opposite the aortic arch. After placement of recurrent laryngeal nerve monitoring equipment (always used), the chest is entered using muscle-sparing techniques, and the lungs are gently retracted anteriorly, the mediastinal pleura opened, revealing the esophagus and/or airway, and the Vagus nerve is gently displaced anteriorly with the trachea. The esophagus is then circumferentially mobilized, staying on the muscular fibers with minimal cautery and frequent use of the recurrent nerve stimulator to avoid recurrent laryngeal nerve injury. Understanding both the normal and alternative courses of the recurrent laryngeal nerves is essential. The spine’s anterior longitudinal ligament is then exposed, and the thoracic duct is protected and rotated into the left mediastinum (when in the right chest). The descending aorta is then exposed, and a descending posterior aortopexy performed if necessary, carefully monitoring the lower body blood pressure with

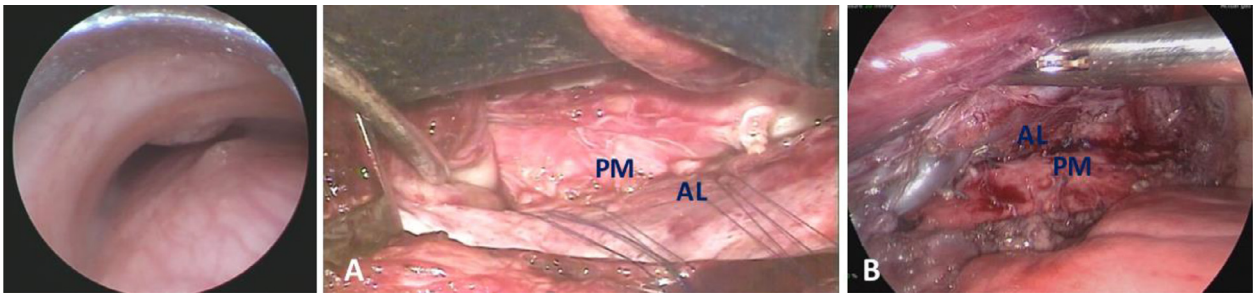

Fig. 5. Posterior tracheopexy to treat severe tracheomalacia via open approach (A) vs. MIS approach (B). Tracheal posterior membrane [PM] is secured to the spinal anterior longitudinal ligament [AL].

an arterial line. The anesthesia team then inserts the flexible bronchoscope, and the posterior tracheobronchopexy is performed by passing autologous pledgeted (often pleura) polypropylene sutures (Prolene®) into but not through the posterior membrane of the trachea and/or main bronchi and securing them to the anterior longitudinal spinal ligament (Fig. 5A). The sites and the direction of the sutures are guided by the intraoperative bronchoscopy.

Symptomatic TBM typically improves following posterior tracheobronchopexy, even in those who have a minor degree of anterior compression (less than 50%). If the patient continues to have persistent symptoms, anterior procedures (aortopexy and anterior tracheobronchopexy) can be considered as a second procedure. In such cases, the posterior tracheobronchopexy provides posterior support of the posterior membrane before pulling the anterior airway wall in the opposite direction with aortopexy and anterior tracheobronchopexy. Anterior tracheobronchopexy employs a similar suturing technique to the posterior pexy procedure to support the anterior wall of the trachea and/or main bronchi to the sternum and anterior chest wall, although we now use pericardium strips as struts between the trachea and the sternum. In both anterior and posterior tracheobronchopexy, suturing is done under direct bronchoscopic guidance to confirm the precision of suture placement, taking care to stay extraluminal, and assess improvement in airway diameter during the tying process. The patients requiring both anterior and posterior procedures typically have bow-shaped airway cartilages with severe (more than 50%) anterior airway compression from great vasculature. In cases with isolated severe anterior vascular compression of the airway or with structural congenital heart disease that needs surgical repair, the anterior work may need to proceed first, and if respiratory symptoms persist, the posterior work is performed later to achieve optimal airway patency. Sometimes we can do both the anterior and posterior work during the same procedure, but this has an increased chance of airway injury to the posterior membrane. At the conclusion of all airway procedures, we do the Munoz maneuver – a negative pressure test of the airway with simultaneous bronchoscopy visualization. Our goal is greater than 50% airway patency at minus 50mmHg suction applied to the airways. In a study by Shieh, reporting on outcomes in our initial 98 patients with symptomatic TBM, significant reduction of symptoms and bronchoscopic evidence of improvement following posterior tracheobronchopexy was reported [22]. Our group has also performed posterior tracheobronchopexy through the MIS approach for the treatment of TBM in select individuals [40,41] (Fig. 5B).

Posterior descending aortopexy

Posterior descending aortopexy has been described to reduce left mainstem bronchus compression from descending aorta [27, 42-44]. A descending aorta located anterior to the spine can cause posterior compression on the mid-portion of the left mainstem bronchus (Fig. 4). In this case, the posterior intrusion may become worse when the bronchus is pexied to the spine's anterior longitudinal ligament during the posterior bronchopexy. In such cases, a posterior descending aortopexy is often performed to secure the descending thoracic aorta to the side of the spine as posteriorly as necessary to relieve the left mainstem bronchus compression between the descending aorta posteriorly and the pulmonary artery anteriorly. This posterior movement of the aorta may necessitate dividing one or more intercostal arteries, and a pre-operative MDCT scan helps to avoid the risk of injury to the artery of Adamkiewicz.

External splinting

External splinting techniques using autologous or synthetic materials have been described in animal models and limited human studies to stabilize collapsible airway [45-48]. Placement of the external splint with synthetic material, such as Silastic meshes or ceramic rings, has raised concerns in terms of long-term effects and complications, including foreign body reaction, infection, and erosion into nearby structures. The need for airway growth in infants and children limits the solutions to resorbable or dilatable possibilities. External splints made from bioresorbable material have been introduced to temporarily provide external airway support while allowing age-proportional growth of the airway with full resorption predicted to occur within 1-3 years. Zoph et al. at the University of Michigan reported trialing the use of 3-D printed, custom-made external splints for the treatment of TBM in critically ill children [49-51]. Using the patient's CT scan, computer software, and laser-based 3-D printing system, a polycaprolactone splint is custom designed and then secured around the affected area to maintain airway support. In a recent case series, 15 patients (median age, 8 months) received 29 splints to treat severe tracheobronchomalacia with significant improvement at a median follow-up of 8.5 months [52].

We found that anterior and posterior tracheobronchopexy have limited utility in cases of transverse tracheal and/or bronchial malformations and that application of an external splint provides better airway support in these conditions, often in combination with the other procedures. Our team at the EAT Center has been utilizing readily available moldable bioresorbable plates (Rapid-Sorb, Synthes CMF) to make customized external splints intraoperatively in patients found to have transverse compression or malformations (scabbard deformities) and circumferential collapse deformities, which require lateral or circumferential support. We pursue direct tracheopexy options for all of our patients as the first-line surgical intervention but find these certain anatomic variants (scabbard or lateral deformities, circumferential collapse) to require additional support from the external splints. Many patients require intraoperative customized surgical solutions and may require combinations of these techniques (Fig 6.). Simultaneous

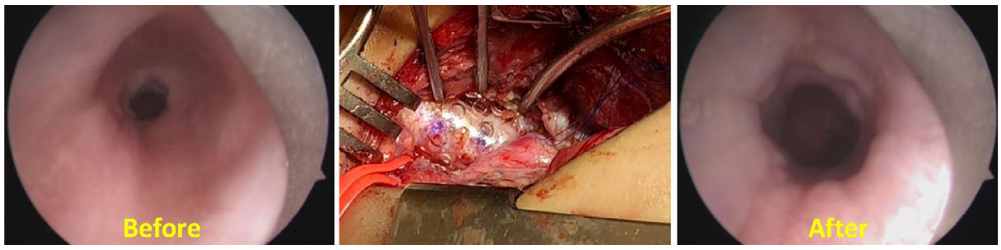

Fig. 6. Customized surgical plan, including placement of anterior external splint (bioresorbable) combined with posterior tracheopexy, to optimize airway opening.

bronchoscopy is used for all cases during repair to guide our choices and help prove that the combination of techniques chosen was effective in optimizing airway opening, and then tested with the Munoz maneuver for effectiveness.

Tracheobronchial resection and reconstruction

Resection of the trachea and/or main bronchus with either end-to-end anastomosis or as a slide tracheobronchoplasty may be indicated in selected patients with some types of short-segment tracheal or bronchial collapse, such as in those with the congenital absence of cartilage or severe cartilage deformation. However, this approach is not widely used in children. TBM rarely affects a short segment of the airway, and pediatric airway typically tolerates less anastomotic tension.

Intraluminal stenting

Endoscopic placement of intraluminal tracheal stents has been attempted using a variety of materials; however, each poses its own potential issues [53-56]. The use of intraluminal stent in children has been limited due to several practical problems and the risk of serious complications, including stent migration or fracture, erosion into nearby structures, the formation of granulation tissue, difficult removal, and the need for additional dilations or stents, especially with patient growth. Currently, this approach has fallen out of favor, but advances in stent development and design can improve the outlook for this as a therapeutic option.

Conclusions

Airway collapse because of dynamic tracheobronchomalacia (TBM), static compression from vascular compression, and/or tracheobronchial deformation are clinically challenging conditions. Patients are best assessed and managed by a multidisciplinary team in centers specializing in complex pediatric airway disorders. Suspicion is made through clinical history and physical examination, diagnosis of location and severity by dynamic 3-phase bronchoscopy, and surgical treatment planning by MDCT and other studies as necessary to completely understand the problems. The treatment plan should be patient-based with a thorough approach to the underlying pathology, clinical concerns, and combined abnormalities. Patients should undergo maximum medical therapy prior to committing to other interventions. For those children considered candidates for surgical intervention and who have failed maximum medical therapy, all other associated conditions, including vascular anomalies, chest wall deformities, mediastinal lesions, or other airway pathologies, should also be considered. Our preference is to correct the airway lesions at the same operation as other comorbidities, if possible, to prevent multiple reoperations with their attendant increased risks. We also strongly advocate for the use of recurrent laryngeal nerve monitoring in all cases of cervical or thoracic surgery to minimize the risks to vocal cord function and laryngeal sensation. Even with treatment, continued surveillance is required to identify any progression or recurrence of symptoms since therapeutic strategies are innovative and not yet standard of care. Similarly, studies that evaluate the effect of our interventions on the patient and caregiver's quality of life are needed to fully grasp the impact of TBM on this challenging patient population.

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