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Innovative Management of Severe Tracheobronchomalacia Using Anterior and Posterior Tracheobronchopexy

Claire Lawlor, MD; Charles Jason Smithers, MD; Thomas Hamilton, MD; Christopher Baird, MD; Reza Rahbar, DDS, MD; Sukgi Choi, MD*; Russell Jennings, MD*

Objectives/Hypothesis: Combined anterior and posterior tracheobronchopexy is a novel surgical approach for the management of severe tracheobronchomalacia (TBM). We present our institutional experience with this procedure. Our objective was to determine the utility and safety of anterior and posterior tracheopexy in the treatment of severe TBM.

Study Design: Retrospective chart review.

Methods: All patients who underwent anterior and posterior tracheopexy from January 2013 to July 2017 were retrospectively reviewed. Charts were reviewed for indications, preoperative work-up, tracheobronchomalacia classification and severity, procedure, associated syndromes, synchronous upper aerodigestive tract lesions, and aberrant thoracic vessels. Main outcomes measured included improvement in respiratory symptoms, successful extubation and/or decannulation, vocal fold immobility, and new tracheotomy placement.

Results: Twenty-five patients underwent anterior and posterior tracheopexy at a mean age of 15.8 months (range, 2–209 months; mean, 31 months if 2 outliers of 206 and 209 months included). Mean length of follow-up was 26.8 months (range, 13–52 months). Indications for surgery included apneic events, ventilator dependence, need for positive pressure ventilation, tracheotomy dependence secondary to TBM, recurrent pneumonia, and exercise intolerance. Many patients had other underlying syndromes and synchronous upper aerodigestive tract lesions (8 VACTERL, 2 CHARGE, 1 trisomy 21, 1 Feingold syndrome, 17 esophageal atresia/tracheoesophageal fistula, 20 cardiac/great vessel anomalies, 1 subglottic stenosis, 1 laryngomalacia, 7 laryngeal cleft). At preoperative bronchoscopy, 21 of 25 patients had >90% collapse of at least one segment of their trachea, and the remaining four had 70% to 90% collapse. Following anterior and posterior tracheopexy, one patient developed new bilateral vocal-fold immobility; one patient with a preoperative left cord paralysis had a new right vocal-fold immobility. Postoperatively, most patients had significant improvement in their respiratory symptoms (21 of 25, 84%) at most recent follow-up. Three patients with preexisting tracheotomy were decannulated; two patients still had a tracheotomy at last follow-up. Two patients required new tracheotomy for bilateral vocal-fold immobility.

Conclusions: Combined anterior and posterior tracheopexy is a promising new technique for the surgical management of severe TBM. Further experience and longer follow-up are needed to validate this contemporary approach and to minimize the risk of recurrent laryngeal nerve injury.

Key Words: tracheomalacia, bronchomalacia, tracheopexy, bronchopexy.

Level of Evidence: 4

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INTRODUCTION

Tracheobronchomalacia (TBM) is a disease state characterized by collapse or compression of the trachea and mainstem bronchi on exhalation, with an incidence of approximately one in 2,500 live births. Tracheal collapse can result from intrinsic weakness of the airway or from extrinsic compression.[1,2] Intrinsic weakness is rare and is associated with weak or soft cartilage. More commonly, the cartilage takes an abnormal shape and/or there is dynamic collapse of the posterior wall of the trachea.[2] Early histopathologic studies of the trachea in children with TBM/tracheoesophageal fistula (TEF) postmortem found a decrease in the normal 4.5:1 ratio of cartilage to posterior membrane muscle, and historically this has been used to characterize TBM.[1] Excessive dynamic airway collapse is a relatively new term specifically used to describe patients with airway narrowing caused by coaptation of a wide, mobile posterior membrane into the airway lumen.[3] The trachea and bronchi can be extrinsically compressed by structures such as normal or aberrant vasculature, thyroid goiters, congenital tumors and cysts, and the spine or sternum.[4] TBM is commonly associated with other syndromes or synchronous airway lesions, such as VACTERL, esophageal atresia (EA)/TEF, and laryngeal and laryngotracheal cleft.[1,2,5–7]

From the Department of Otolaryngology (C.L.), Children's National Medical Center, Washington, DC; and the Department of Surgery (C.J.S., T.H., R.J.), Department of Cardiac Surgery (C.B.), and Department of Otolaryngology and Communication Enhancement (R.R., S.C.), Boston Children's Hospital, Boston, Massachusetts, U.S.A.

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Send correspondence to Sukgi S. Choi, MD, Department of Otolaryngology, Boston Children's Hospital, 300 Longwood Ave BCH3129, Boston, MA 02115. E-mail: sukgi.choi@childrens.harvard.edu

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TBM usually presents within the first 2 to 3 months of life.[2,8] The most common presentation includes barking cough and biphasic or expiratory stridor. Recurrent or prolonged respiratory infections, feeding difficulties, increased work of breathing, wheezing, cyanosis, and/or apnea may also be present.[2,9] TBM can be divided into mild, moderate, and severe forms. Mild forms can present as recurrent or prolonged respiratory infections. Mild TBM may have a self-limited course, which may improve as the child's airway grows, sometimes by age 2 years, and therefore may go undiagnosed.[10] Moderate and severe cases present with expiratory stridor, barky cough, recurrent and prolonged infections, and apneic apparent life-threatening event (ALTE)/brief resolved unexplained event (BRUE) episodes.[11] The mortality of severe TBM may be as high as 80%.[1] Direct bronchoscopy is the gold standard for diagnosis of TBM, with multidetector computed tomography (MDCT) serving as an adjunct source of information.[12] Historically, a dynamic reduction in the tracheal diameter of more than 50% associated with classic symptoms was used as the threshold to diagnosis of TBM on bronchoscopy.[2]

Primary treatment modalities for severe TBM failing expectant management have included tracheotomy often with positive pressure ventilation (PPV) to "stent" the collapsing airway, tracheal stents to hold the airway open, and anterior aortopexy to suspend the vessel and alleviate much of the external vascular compression on the anterior trachea.[1,13] Great strides have been made within the past 5 to 10 years to advance the surgical management of TBM. Posterior tracheopexy, which tacks the posterior membranous wall of the trachea to the spine, has demonstrated success in correcting posterior membrane intrusion.[11,14–16] However, a subset of patients may benefit from both anterior aortopexy, anterior tracheopexy, and posterior tracheobronchopexy. We present a cohort of patients requiring this novel surgical combination and review their operative criteria and surgical outcomes.

MATERIALS AND METHODS

We retrospectively reviewed all patients who underwent anterior and posterior tracheobronchopexy at Boston Children's Hospital from January 2013 to July 2017 under an approved institutional review board protocol (IRB-P00026671). All patients who underwent anterior and posterior tracheobronchopexy during this time were included. Anterior and posterior tracheobronchopexy procedures were performed by the Esophageal and Airway Treatment team at Boston Children's Hospital, which consists of three primary pediatric surgeons, one pediatric cardiothoracic surgeon, one pediatric pulmonologist, two pediatric gastroenterologists, and two pediatric otolaryngologists. All pre- and postoperative dynamic bronchoscopies were performed by the pediatric surgeons and/or pediatric otolaryngologists.

A review of patient records was conducted, and data were collected, including patient age at the time of surgery, gender, presenting symptoms, comorbid conditions, synchronous direct laryngotracheobronchoscopy findings, surgical procedures, time between operations, total number of operations, resolution of preoperative respiratory symptoms, complications of anterior/posterior tracheobronchopexy, and length of follow-up. Main outcomes measured were patient specific and included improvement in apnea, frequency of respiratory illnesses, exercise intolerance, ventilator or oxygen weaning, extubation and/or decannulation, vocal-fold immobility, and new tracheotomy placement.

Decision Making for Treatment of TBM

An algorithm for the use of anterior and posterior tracheopexy in the treatment of severe TBM is shown in Figure 1. Work-up for TBM is based on patient signs and symptoms, including barky cough, recurrent respiratory infections, prolonged respiratory infections, exercise intolerance, development of bronchiectasis, and ALTE/BRUE.

The diagnosis of TBM is made during dynamic, three-phase bronchoscopy, which is the preferred method of the pediatric surgeons at our institution, who participated in or performed the bronchoscopies on this cohort.[14] The first phase of the dynamic bronchoscopy is performed using a ventilating bronchoscope with the patient taking spontaneous, shallow breaths. This allows for evaluation of static compression of the trachea and bronchi, as well as identification of synchronous airway anomalies. In the

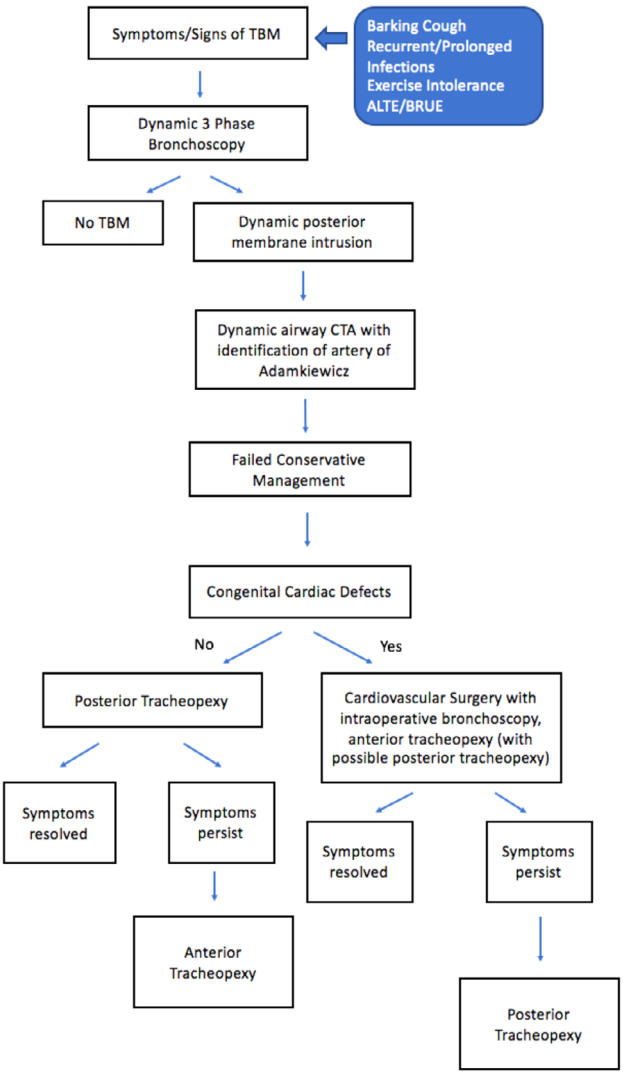

Fig. 1. Management algorithm for anterior/posterior tracheopexy. ALTE = apparent life-threatening event; BRUE = brief resolved unexplained event; CTA = computed tomography angiography; TBM = tracheobronchomalacia.

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second phase of bronchoscopy, the aim is to see the patient's airway during valsalva or cough to assess maximum dynamic motion and collapse during expiration (Fig. 2). This is achieved by lightening the anesthetic until the patient becomes sensitive to stimulation in the airway (either via the bronchoscope itself or a flexible suction catheter) and begins to cough. In the final phase of bronchoscopy, all secretions are removed, and the airway is examined while the patient is sedated with the airway distended to 40 cm H_2O (30 cm H_2O in neonates). This procedure facilitates evaluation of the posterior membrane for TEF and tracheal diverticulum. Patients who are candidates for surgery based on clinical findings and dynamic, three-phase bronchoscopy then undergo a dynamic airway MDCT angiogram with artery of Adamkiewicz protocol (Fig. 3) to understand the locations of the mediastinal structures, identify vascular or mass compression, and plan surgical intervention.

Conservative management is first attempted for all patients diagnosed with TBM. Medical therapies include hypertonic saline nebulizer treatments to thin airway secretions and facilitate clearance, inhaled ipratropium bromide to decrease secretions, and chest physiotherapy. Inhaled low-dose corticosteroids and antibiotics to decrease airway inflammation are minimized and used only during active infections. Continuous positive airway pressure (CPAP) and bilevel positive airway pressure are noninvasive treatments for TBM and can be delivered via face masks, endotracheal tubes, or tracheotomy tubes. However, dependence on PPV and/or tracheotomy, recurrent respiratory infections precipitating multiple hospitalizations, ALTE/BRUE, and, more often in older patients, exercise intolerance that persists despite conservative management will prompt surgical management. The duration of conservative management attempted is dependent on individual presentation and severity of symptoms and is usually 1 month at minimum.

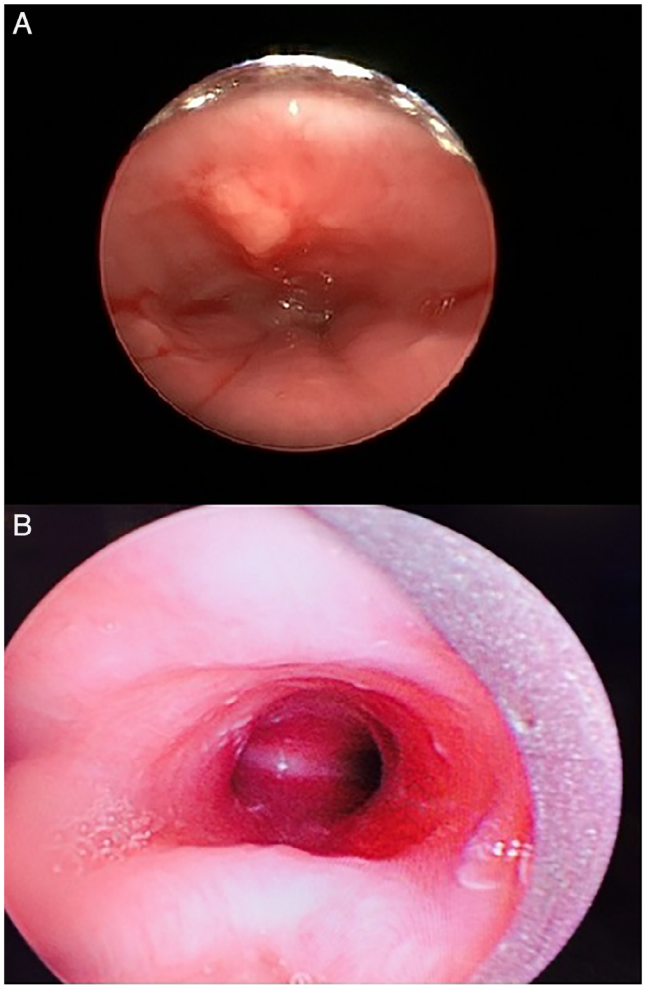

Fig. 2. (A) Severe tracheobronchomalacia with anterior compression and posterior intrusion on dynamic bronchoscopy. (B) Airway patency after anterior and posterior tracheopexy on dynamic bronchoscopy during Valsalva or cough. (Photos are from different patients.)

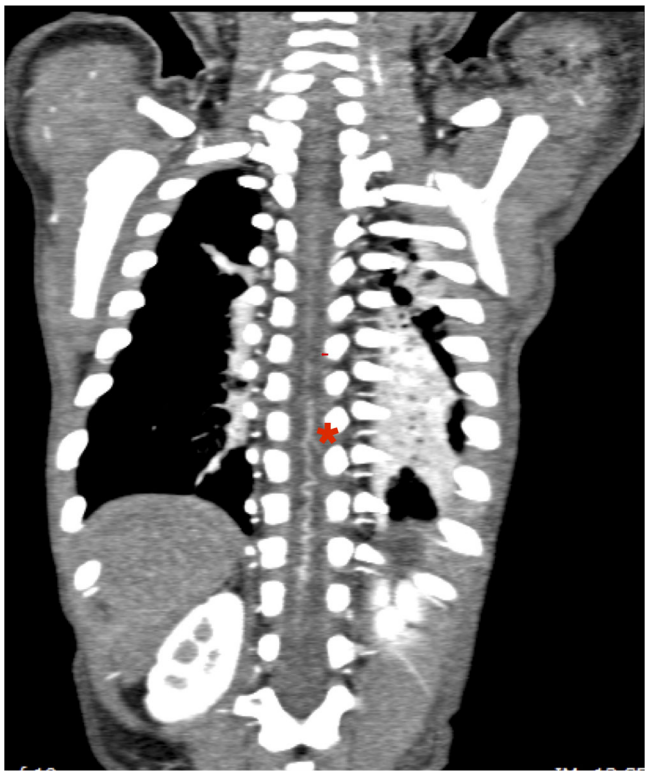

Fig. 3. Dynamic airway computed tomography angiogram with artery of Adamkiewicz (AoA) protocol. The AoA is the primary blood supply to the thoracolumbar spine; the aorta supplies the AOA, which feeds the anterior spinal artery. Oblique coronal 1.3- to 1.5-mm-thick maximum intensity projections are reconstructed from the phase where the AoA is identified. The AoA is identified as a classic "hairpin loop" (red asterisk) within the spinal canal as it arises from an intercostal artery and feeds into the anterior spinal artery.

It is important to note that we also perform anterior aortopexy in isolation for TBM at our institution; this decision is made based on individual patient presentation and bronchoscopic findings. In patients with significant posterior membrane intrusion without need for cardiac surgery, we first support the posterior membrane using posterior tracheobronchopexy. Typically, the posterior tracheobronchopexy is performed through the right fourth intercostal space (in those patients with a left aortic arch), retracting the lungs anteriorly and rotating the esophagus laterally. Posterior tracheopexy is performed by passing autologous pledgeted polypropylene sutures into but not through the posterior tracheal membrane and securing them to the anterior longitudinal ligament of the spine. The pledgets are made from pleura, pericardium, or fibrous scar tissue as opposed to synthetic material and protect the delicate structures from suture damage. The sutures are placed under intraoperative flexible bronchoscopic visualization, which allows the surgeon to assess the result of each suture on the airway while avoiding suture placement into the lumen of the trachea. The number and placement of

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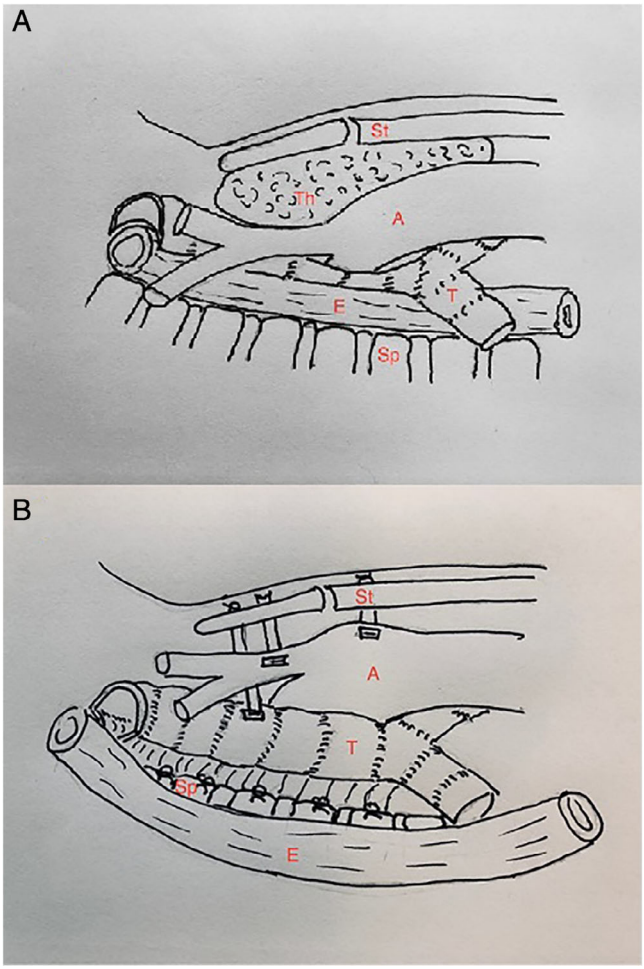

Fig. 4. Illustrations of thoracic structures before (A) and after (B) anterior and posterior tracheopexy. A = aorta; E = esophagus; Sp = spine; St = sternum; T = trachea; Th = Thymus.

sutures is guided by the patient's anatomy; a typical range is six to 24 interrupted sutures. At the end of the procedure, negative pressure is applied to the airway with flexible bronchoscopy to visualize the airway and confirm noncollapsibility. This technique significantly improves or resolves symptoms in approximately 90% of our patients with severe TBM, and they do not require anterior aortopexy/tracheopexy.[15]

In those patients who do not have complete resolution of symptoms and continue to have demonstrable TBM on dynamic, three-phase bronchoscopy, an anterior approach is utilized to perform anterior tracheopexy. A mini-sternotomy allows approach to all the structures in the mediastinum. Initially, the thymus is removed to improve access in the anterior mediastinum. With intraoperative flexible bronchoscopy used to guide the surgeon, ascending aortopexy is performed by opening the pericardium and suspending the ascending aorta from the sternum. This may be all that is needed to improve the airway. The innominate artery may require suspension as well. Often, these maneuvers are not enough to adequately open the airway due to cartilage malformation or compression. In those patients, the anterior wall of the trachea is exposed and direct anterior tracheopexy is performed, supporting the trachea to the sternum. Due to the prior posterior membrane fixation after the posterior tracheopexy, usually very little movement of the anterior trachea is needed to optimize airway anatomy. After sternal closure, the airway is tested under negative pressure with flexible bronchoscopy to assess the effects of the correction. Illustration of thoracic structures before and after anterior and posterior tracheopexy can be seen in Figure 4.

TABLE I.
Surgical Indications for Anterior and Posterior Tracheopexy, Often Multifactorial.

Surgical Indication	N (%)
Recurrent respiratory infections	12 (48)
Ventilator/PPV dependence	11 (44)
Increased work of breathing/respiratory distress	8 (32)
ALTE/BRUE	5 (20)
Tracheotomy dependence	5 (20)
Exercise intolerance	2 (8)

ALTE = apparent life-threatening event; BRUE = brief resolved unexplained event; PPV = positive pressure ventilation.

In patients who require cardiac surgery on bypass, the cardiac and tracheal surgeries can be combined into one operation guided by the intraoperative bronchoscopy. A sternotomy approach affords excellent exposure for cardiac surgery and anterior aortopexy. The anterior tracheopexy sutures are guided by intraoperative bronchoscopy to optimize the anterior airway support. Unfortunately, access to the posterior trachea via a sternotomy is very difficult. The child may require cardiac bypass with the heart decompressed. The trachea can then be rotated 90° to place the posterior tracheopexy sutures. The sutures cannot easily be placed under bronchoscopic guidance, although suture placement can be verified after the trachea is rotated back to its anatomical position. Mobilizing the esophagus for access to the spine from the anterior approach is challenging and places the vagus and recurrent laryngeal nerves (RLNs) at greater risk of injury than does a thoracoscopic or thoracotomy approach.

The rationale for performing the anterior and posterior tracheopexy versus just anterior tracheopexy at the time of cardiac surgery is related to the complexities associated with exposure. At present, if preoperative dynamic bronchoscopy demonstrates significant posterior membrane intrusion, we will perform anterior and posterior tracheopexy at the time of cardiac surgery. Occasionally, anterior tracheopexy will be performed at the time of cardiac

TABLE II.
Syndromes and Anatomical Anomalies Associated With Patients Undergoing Anterior and Posterior Tracheopexy.

	No.
Syndromes	
VACTERL	8
CHARGE	2
Trisomy 21	1
Feingold	1
Synchronous anomalies	
Cardiac/great vessel	20
Subglottic stenosis	1
Laryngomalacia	1
Laryngeal cleft	
Type 1	2
Type 2	4
Type 3	1
Unilateral vocal-fold immobility	3

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TABLE III.
Patient Characteristics, Comorbidities, Surgical Indications, Procedures, and Outcomes.

Patient	Age (Mo)	Comorbidities	Prior Procedures	Surgical Indications	Preoperative TVC Mobility	TBM Classification	Procedure 1	Residual Symptoms	Procedure 2	Operative Interval	Other Procedures	Outcomes	Postoperative TVC Mobility
1	6	CHARGE, type II laryngeal cleft, EA/TEF, tracheal diverticulum, coarctation of aorta, bicuspid aortic valve, VSD, large PDA, BPD	EA/TEF repair; repair of coarctation of the aorta, PDA ligation	Prolonged intubation, extubation to BiPAP 19/13, desaturations/bradycardia with agitation despite PPV, recurrent pneumonia	Mobile bilaterally	80% collapse of midtrachea, complete collapse at distal trachea; L mainstem bronchus >50% collapse	Posterior tracheopexy, EA/TEF repair, resection of tracheal diverticulum	Persistent CPAP	Anterior tracheopexy, anterior aortopexy, and innominate artery pexy	25	Tracheotomy; type II laryngeal cleft repair	50%–70% collapse L mainstem bronchus, still on CPAP 6, no further aspiration or desaturations/bradycardia with agitation	Immobile bilaterally
2	56	VACTERL, type II laryngeal cleft, EA/TEF, tracheal diverticulum, aberrant R subclavian artery	EA/TEF repair; tracheotomy	Trach dependence, recurrent respiratory infection	Mobile bilaterally	Complete collapse of mid- and distal trachea; L mainstem bronchomalacia >50%	Posterior tracheopexy, resection of tracheal diverticulum, repair of aberrant subclavian artery	Persistent CPAP	Anterior tracheopexy, anterior aortopexy	359	Type II laryngeal cleft	<50% collapse in all segments on DLB, decannulated	Mobile bilaterally
3	209	Double aortic arch	Division of double aortic arch; suspension of aortic arch	Dyspnea on exertion/exercise intolerance, recurrent infections	Mobile bilaterally	>70% collapse distal trachea and R mainstem bronchus, external compression	Anterior tracheopexy, posterior tracheopexy, anterior aortopexy, R mainstem bronchopexy	NA	NA	N/A	N/A	Improved exercise intolerance, resolved frequent pneumonias	Mobile bilaterally
4	5	Double aortic arch, EA/TEF, BPD	EA/TEF repair; double aortic arch repair	BiPAP settings 14/8	L immobile before	Midtrachea complete collapse	Anterior tracheopexy, posterior tracheopexy, tracheal diverticulum resection, repair of double aortic arch	NA	NA	NA	Tracheotomy	Weaned to CPAP PEEP 6	New R TVC immobility, immobile bilaterally
5	12	VACTERL, EA/TEF	None	Repeated intubations for respiratory failure	Mobile bilaterally	Complete collapse at midtrachea, innominate compression	Posterior tracheopexy, TEF repair, tracheal diverticulum repair	Recurrent respiratory infections	Revision posterior tracheopexy, anterior tracheopexy, anterior aortopexy	284	NA	No reintubations or respiratory distress, recurrent severe/prolonged respiratory infections resolved	Mobile bilaterally
6	10	VACTERL, EA, dextrocardia, ASD	None	Ventilator dependence, intubated	Unknown	>90% collapse at proximal/midtrachea	Posterior tracheopexy, EA repair, excision of tracheal diverticulum	Respiratory distress	Anterior aortopexy, anterior tracheopexy	119	NA	Doing well, on room air	Initially bilateral weakness, improved
7	13	Hypoplastic aortic arch, VSD, severe distal bronchomalacia	Hypoplastic aortic arch/VSD repair; tracheotomy	Trach dependence, severe desaturations/cyanosis, PEEP 12	Mobile bilaterally	Complete collapse of mid/distal trachea, bronchomalacia	Posterior tracheopexy	Profound desaturations with agitation despite PEEP	Anterior tracheopexy	15	NA	Persistent severe distal bronchomalacia, PEEP still 12, still needs trach	Mobile bilaterally
8	16	EA/TEF, ASD, L-sided spit fistula, Feingold, type I cleft	EA/TEF repair w/ L spit fistula	Apneic episodes/ALTEs	Mobile bilaterally	>80 collapse distal trachea, complete collapse bilateral mainstem bronchi	Anterior tracheopexy, posterior tracheopexy, anterior aortopexy, RPA pexy, resection of	NA	NA	NA	NA	No further apneic episodes, doing well	Mobile bilaterally

TABLE III.
(Continued)

Patient	Age (Mo)	Comorbidities	Prior Procedures	Surgical Indications	Preoperative TVC Mobility	TBM Classification	Procedure 1	Residual Symptoms	Procedure 2	Operative Interval	Other Procedures	Outcomes	Postoperative TVC Mobility
							tracheal diverticulum, ASD						
9	203	EA/TEF, hypoplastic R lung, hiatal hernia, dextrocardia, VACTERL	Tracheotomy, EA/TEF repair, hiatal hernia repair	Trach dependence, CPAP	Mobile bilaterally	Complete collapse in mid/distal trachea	Anterior tracheopexy, anterior aortopexy, posterior tracheopexy, resection of tracheal diverticulum, R pneumonectomy	NA	NA	NA	NA	Decannulated, on room air, doing well	Mobile bilaterally
10	40	EA/TEF, VACTERL, VSD, PDA	EA repair, PDA ligation	ALTEs, recurrent respiratory infections	Mobile bilaterally	Complete collapse of the distal trachea	Anterior tracheopexy, anterior aortopexy, VSD/PFO repair	Continued recurrent respiratory infections	Posterior tracheopexy, resection of tracheal diverticulum	1,138	NA	No further severe/prolonged respiratory infections, ALTEs resolved	Mobile bilaterally
11	14	EA	EA repair	Respiratory distress, recurrent respiratory infections	Mobile bilaterally	Complete collapse of distal trachea, anterior compression	Anterior tracheopexy, anterior aortopexy	Persistent difficulty breathing, recurrent infections	Posterior tracheopexy	70	NA	No further severe/prolonged respiratory infections, increased WOB resolved	Mobile bilaterally
12	6	EA/TEF, SGS, type II laryngeal cleft	EA/TEF repair, tracheotomy, type II laryngeal cleft repair	Trach dependence, recurrent respiratory infection	Mobile bilaterally	Complete collapse of mid/distal trachea, bilateral mainstem bronchi	Posterior tracheopexy, resection of tracheal diverticulum	Continued trach and CPAP dependence	Anterior tracheopexy, anterior aortopexy	174	Type II laryngeal cleft repair, LTR	Decannulated, on room air, no further severe/prolonged respiratory infections	Mobile bilaterally
13	9	EA, tracheal diverticulum, pHTN, ASD, VACTERL	None	Recurrent infections, respiratory distress, need for CPAP	Mobile bilaterally	Complete collapse of mid/distal trachea, bilateral mainstem bronchi	Posterior tracheopexy, EA repair, excision of tracheal diverticulum	Residual respiratory distress, infections	Anterior tracheopexy, anterior aortopexy	134	NA	On room air, no further severe/prolonged respiratory infections	Mobile bilaterally
14	5	EA, tracheal diverticulum, VACTERL	EA repair	Ventilator dependence, intubated	Unknown	Complete collapse of midtrachea, L mainstem collapse; severe anterior compression	Posterior tracheopexy, EA repair, excision of tracheal diverticulum	Failed extubation	Anterior tracheopexy, anterior aortopexy	61	NA	Extubated to room air, doing well	Mobile bilaterally
15	11	R aortic arch, circumflex aorta	None	Recurrent respiratory infections, respiratory distress	Mobile bilaterally	80% collapse of distal trachea, severe bilateral mainstem bronchi collapse	Anterior tracheopexy, posterior tracheopexy, aortopexy, aortic arch reconstruction	NA	NA	NA	NA	No further severe/prolonged respiratory infections, increased WOB resolved	Mobile bilaterally
16	3	TOF, aberrant left subclavian, pulmonary valve regurgitation and stenosis with dual LAD of the R aortic arch	None	CPAP dependent, respiratory distress, cyanosis	Unknown	Complete collapse at carina, mainstem bilaterally	Anterior tracheopexy, posterior tracheopexy, aortoplasty, closure of VSD	NA	NA	NA	NA	Off positive pressure ventilation, on room air, doing well	Mobile bilaterally
17	3	R aortic arch, large VSD, laryngomalacia	None	Desaturations, cyanosis, respiratory distress	Mobile bilaterally	Complete collapse at mid/distal trachea and L mainstem bronchus	Anterior tracheopexy, posterior tracheopexy, aortic arch reconstruction, VSD/ASD closure	Persistent CPAP	Anterior tracheopexy	64	Supraglottoplasty	Off CPAP, doing well, on room air	Mobile bilaterally
18	39	VACTEREL, ASD, type II laryngeal cleft, anoxic brain injury with autonomic instability, seizure disorder, chronic hypoxemic respiratory failure	Tracheotomy, ASD repair	Trach dependence, frequent desaturations to 10%, cyanotic episodes, ALTEs	Mobile bilaterally	Complete collapse of mid/distal trachea	Posterior tracheopexy	Continued anterior collapse, desaturations	Anterior tracheopexy, anterior aortopexy	15	NA	**S**till has trach, still requires intermittent CPAP, ALTEs and severe desaturations resolved	Mobile bilaterally

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19	2	TOF, absent pulmonary valve	None	Intubated at birth, severe airway compression	Unknown	Complete collapse of distal trachea, R mainstem bronchus	Anterior tracheopexy, TOF repair, resection of branch main PA, resection of ascending aorta, LeCompte procedure	Continued high-flow O_2 requirements, airway compression	Posterior tracheopexy	72	NA	On room air, doing well	Mobile bilaterally
20	9	EA/TEF, PDA, type III laryngeal cleft, L TVC immobility	EA/TEF repair x2, PDA ligation, laryngeal cleft repair	ALTE, recurrent respiratory infections, ventilator dependence	L immobile before	Complete collapse mid/distal trachea	Anterior tracheopexy, posterior tracheopexy, aortopexy, repair tracheal diverticulum, resection mediastinal cyst	NA	NA	NA	NA	On room air, no further severe/prolonged respiratory infections or ALTEs	L immobile before, R mobile
21	16	EA/TEF, type I laryngeal cleft, VACTERL	EA repair	ALTE, cyanosis with feeding	Unknown	Complete collapse midtrachea	Posterior tracheopexy, EA/TEF repair, removal of tracheal FB	Recurrent respiratory infections	Anterior aortopexy, anterior tracheopexy	67	Type I laryngeal cleft repair	On room air, no further severe/prolonged respiratory infections, no further ALTEs	Mobile bilaterally
22	20	EA/TEF, PFO, bifurcating R innominate artery	EA/TEF repair	Recurrent respiratory infections, respiratory distress	Mobile bilaterally	Complete collapse mid/distal trachea	Posterior tracheopexy, resection of tracheal diverticulum	Recurrent respiratory infections	Aortopexy, anterior tracheopexy	207	NA	On room air, doing well, no further infections	Mobile bilaterally
23	44	Unbalanced complete AV canal defect, EA/TEF; trisomy 21	L pulmonary artery plasty, L AV valve plasty, closure of L AV canal; Norwood; Glenn; EA/TEF repair	Recurrent respiratory infections	L immobile before	Complete collapse mid/distal trachea, L mainstem bronchus	Posterior tracheopexy, anterior tracheopexy, posterior aortopexy, anterior aortopexy, resection tracheal diverticulum, aortic coarc patch repair, pulmonary artery reconstruction	NA	NA	NA	NA	On room air, no further hospitalizations for respiratory infections	L immobile before, R mobile
24	18	Circumflex aortic arch	None	Recurrent respiratory infections, exercise intolerance	Mobile bilaterally	>80% narrowing distal trachea w/ posterior pulsatile mass	Posterior tracheopexy, anterior tracheopexy, aortic uncrossing	NA	NA	NA	NA	No further severe/prolonged respiratory infections	Mobile bilaterally
25	6	EA/TEF, ASD/VSD, PV stenosis, CHARGE, intestinal malrotation, diaphragmatic hernia	None	Respiratory distress requiring intubation	Mobile bilaterally	>80% collapse at distal trachea	Posterior tracheopexy, EA repair Foker 1	Respiratory distress, recurrent respiratory infections	Anterior tracheopexy, anterior aortopexy, posterior aortopexy, ASD repair, revision posterior tracheopexy, repair diaphragmatic hernia	140	NA	On room air, no further hospitalizations for respiratory infections	Mobile bilaterally

ALTE = apneic apparent life-threatening event; ASD = atrial septal defect; AV = atrioventricular; BPD = bronchopulmonary dysplasia; BiPAP = bilevel positive airway pressure; CPAP = continuous positive airway pressure; DLB = diagnostic laryngoscopy and bronchoscopy; EA = esophageal atresia; FB = flexible bronchoscopy; L = left; LAD = large airway disease; LTR = laryngotracheal reconstruction; NA = not available; PA = pulmonary artery; PDA = patent ductus arteriosus; PEEP = positive end-expiratory pressure; PFO = patent foramen ovale; pHTN = pulmonary hypertension; PPV = positive pressure ventilation; PV = pulmonary valve; R = right; SGS = subglottic stenosis; TBM = tracheobronchomalacia; TEF = tracheoesophageal fistula; TOF = tetralogy of Fallot; Trach = tracheostomy; TVC = true vocal cord; VSD = ventricular septal defect; WOB = work of breathing.

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surgery, and posterior tracheopexy will follow at a later date for persistent symptoms and signs of TBM on dynamic bronchoscopy.

At our institution, close to 300 patients have undergone posterior tracheopexy for severe TBM. The 25 patients reported in this series represent the only patients to have undergone both anterior and posterior tracheopexy.

RESULTS

Twenty-five patients underwent anterior and posterior tracheopexy at a mean age of 15.8 months (range, 2–209 months; mean, 31 months if 2 outliers of 206 and 209 months are included). Mean length of follow-up was 26.8 months (range, 14–52 months). Surgical indications were often multifactorial and are presented in Table I. Many patients had other underlying syndromes and synchronous upper aerodigestive tract lesions (Table II). Patient characteristics, including comorbidities, preoperative indications, procedures, and outcomes can be found in Table III. Three patients with preoperative vocal-fold immobilities were identified, although these were likely related to prior surgeries at outside institutions. The status of vocal-fold mobility was unknown preoperatively in five patients.

All patients underwent pre- and postoperative bronchoscopy. At preoperative bronchoscopy, 21 of 25 patients had a complete collapse of at least one segment of their trachea, and the remaining four had 70% to 90% collapse. Anterior and posterior tracheopexy was performed during the same operation in nine of 25 patients (36%) during cardiac surgery. Posterior tracheopexy was performed first, followed by anterior tracheopexy at a later date in 12 of 25 patients (48%). Anterior tracheopexy was performed during cardiac surgery and preceding posterior tracheopexy in four of 25 patients (16%). When the two procedures were not performed during the same operation, the mean operative interval was 184 days (range, 15–1,138 days). Concurrent aortopexy was performed in 19 of 25 patients (76%). All six patients that did not undergo aortopexy had aortic arch anomalies. Two of these patients had simultaneous thoracotomy with posterior tracheopexy and then sternotomy with anterior tracheopexy, which was reserved for those patients who had insufficient improvement on intraoperative bronchoscopy after posterior tracheopexy.

Postoperatively, most patients had significant improvement in their preoperative respiratory symptoms (21 of 25, 84%) at most recent follow-up; specific outcomes for each patient can be found in Table III. Decannulation was achieved in three patients with preexisting tracheotomy; two patients still had a tracheotomy at last follow-up for continued airway small collapse (lobar and segmental), chronic lung disease, and ventilator requirements related to complications of anoxic brain injury. Following anterior and posterior tracheopexy, one patient developed new bilateral vocal-fold immobility; one patient with a preoperative left cord paralysis had a new right vocal-fold immobility. Both patients with new vocal-fold immobilities underwent simultaneous, challenging EA/TEF revisions after a primary repair at an outside institution; the patient that developed a new right vocal-fold immobility also underwent simultaneous double aortic arch repair. Postoperatively, new tracheotomy was performed in two patients for bilateral vocal-fold immobility; both still needed intermittent PPV, although the settings were improved. One patient developed temporary vocal-fold immobility that resolved without intervention. One patient had a chyle leak that was treated nonoperatively. There were no postoperative deaths.

Identification and repair of synchronous airway lesions was performed by pediatric otolaryngologists. Three patients had true vocal-fold weaknesses diagnosed preoperatively, all unilateral left cord, presumed to have been caused by prior cardiac or EA surgeries. The one patient with synchronous laryngomalacia required a supraglottoplasty. Laryngeal cleft was identified but not repaired in two of seven patients (29%; 1 type 1, 1 type 2). Cleft repairs were performed in five of seven patients diagnosed with laryngeal cleft (50%; 1 type 1, 3 type 2, and 1 type 3). All cleft repairs were performed endoscopically. One patient had concurrent subglottic stenosis; one required laryngotracheal reconstruction before decannulation following surgery for TBM.

DISCUSSION

It is important that the otolaryngologist is familiar with the diagnosis and treatment of TBM, as presenting complaints of cough, expiratory stridor, cyanotic/apneic episodes, and recurrent or prolonged respiratory infections often prompt otolaryngology referrals. Direct tracheobronchoscopy is the gold standard for diagnosis of TBM, but we advocate for the use of dynamic three-phase bronchoscopy to facilitate classification and to diagnose dynamic collapse, TEF, or tracheal diverticula that may be missed during a static shallow-breathing evaluation. TBM is often associated with synchronous airway lesions and should be considered during direct laryngoscopies and bronchoscopies performed on children with airway symptoms.[1,2,5–7] The strongest association appears to exist between TBM and EA/TEF and tracheal diverticulum.[2,15] Synchronous laryngomalacia, laryngeal cleft, vocal-fold immobility, and subglottic stenosis have also been reported.[5–7]

Otolaryngologists are critical members of multidisciplinary teams that care for patients with TBM. The role of the otolaryngologist includes performing flexible laryngoscopies to evaluate pre- and postoperative vocal-fold mobility and direct laryngoscopies and bronchoscopies to diagnose TBM and synchronous lesions and functioning as a member of the surgical team when addressing TBM. When pediatric otolaryngologists diagnose TBM, it is important to be able to discuss the treatment options available for the patient. The association of synchronous airway disease in our cohort, including subglottic stenosis and laryngomalacia, underscores the importance of involving otolaryngologists in the care of these complex patients.

A Cochrane review of interventions for pediatric TBM, performed before these techniques were developed, showed a dearth of quality evidence to support any means of treatment.[17] Today's surgeons treating TBM have many new techniques in their arsenal, and each patient's unique presentation should guide treatment and surgical approach. We compare our innovative procedures to the most commonly reported techniques. When posterior tracheopexy was first described by our group, of the nine

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patients that had preoperative tracheotomies, only two could be decannulated after posterior tracheopexy. Statistically significant improvements in respiratory symptoms were reported without data.[15] However, with more experience, we have documented significant improvement in TBM with posterior tracheal procedures in isolation.[18,19] In 2017, Morrison et al. described their 10-year experience treating severe TBM with aortopexies, intraluminal stents, and tracheotomies; improvement in respiratory symptoms was reported in 75%, 81%, and 67% of patients, respectively. Severe complications, including chylothorax, tracheostomy plugging, and stent fracture, occurred in 12%, 24%, and 33%, respectively.[20] Anterior aortopexy, a procedure commonly utilized to treat TBM, has documented success rates from 60% to 100% and is also not without risk. Complications can occur in as many as 16% of patients, including pneumothorax, pericardial effusion, phrenic nerve injury, and death.[21,22] Injury to the RLN, despite an incidence of approximately 1% after thoracic surgery, is not commonly documented in the TBM literature.[23]

In this article, we describe a novel surgical technique in the management of TBM: combined anterior and posterior tracheobronchopexy. In our 25-patient cohort, 84% of patients experienced improvement in their preoperative respiratory symptoms. Patients with severe TBM that requires anterior and posterior tracheopexy are usually complex and may have comorbid respiratory diseases, such has bronchomalacia, bronchopulmonary dysplasia, and chronic lung disease. These illnesses can confound postoperative evaluation of improvement in respiratory symptoms. Thus, we considered our surgeries successful if our patients had significant improvement or resolution of their surgical indications, such as resolution in apneic spells or weaning off PPV. Of the five patients who required preoperative tracheotomy for respiratory support, three were decannulated during the period of documented follow-up. One of those patients had severe subglottic stenosis and required a laryngotracheal reconstruction prior to decannulation. Two patients were unable to be decannulated; both continued to require PPV. Both of those patients had lower ventilator pressures, but severe chronic lung disease or distal small airway bronchomalacia required continued ventilator support. Symptoms did not improve in four of 25 (16%) patients when compared to their preoperative complaints. Two of the patients considered failures were those unable to be decannulated; it should be noted that their chronic lung disease may confound ventilator weaning. The other two patients were those who had tracheotomy placed after anterior and posterior tracheopexy for bilateral true vocal fold immobility; both of those patients continued to require intermittent PPV.

Anterior and posterior tracheopexy is an extensive surgery and is not without morbidity. All patients in our cohort are still living. Due to the extensive anterior and posterior tracheal dissection, a vexing potential postoperative complication is new unilateral or bilateral true vocal fold immobility or paralysis that may significantly worsen a tenuous patient's respiratory status. Unfortunately, despite many of our patients having had prior surgery, flexible laryngoscopy to assess vocal fold mobility preoperatively was not performed on all patients to document normal preoperative function. Postoperative new vocal fold immobility was identified in two of our patients (1 bilateral and 1 unilateral coupled with preoperative contralateral paralysis, resulting in bilateral immobility), resulting in new postoperative tracheotomy placement. This underscores the important role of otolaryngology in the preoperative planning, specifically to perform preoperative flexible laryngoscopic evaluation and direct laryngoscopy and bronchoscopy to assess for synchronous airway lesions.

Our patient with postoperative temporary vocal fold paresis underscores the need for close otolaryngology follow-up in these patients. Both patients with new vocal fold immobility had complex congenital anomalies. Those patients underwent simultaneous revision EA/TEF repair after a prior repair had been performed at an outside institution. Revision EA/TEF repairs can be very technically challenging, with extensive infection, inflammation, and scar in the tracheoesophageal grooves. One of the patients also had a concurrent double aortic arch repair. These procedures also place the RLN at risk for injury. Complex procedures in the neck and mediastinum do place the RLN at risk for temporary or permanent injury. In retrospect, it is impossible to identify where and when during those technically challenging multilevel surgeries the RLNs were injured. Our team is actively pursuing methods to protect and/or intraoperatively monitor the RLNs during these procedures. We also acknowledge that this potential morbidity is crucial to discuss during the consent process prior to performing these procedures. Otolaryngologists play a vital role in the management of this complication, with tracheotomies and vocal fold medializations as indicated.

Limitations of this report include the small size of the cohort. As this procedure is relatively new, our longitudinal data collection on patients is relatively short. Outcome measures such as improvement in respiratory symptoms can be subjective and dependent on length of follow-up, although resolution of need for PPV or supplemental oxygen, repeated hospitalizations, tracheotomy decannulation, and CPAP settings do provide some objective data. Further investigation into means of protecting the RLN is vital to the success of these technically challenging procedures. This report is not meant to be a comment on the efficacy of anterior or posterior tracheopexy procedures in isolation; outcomes of posterior tracheopexy have been published and are continually reassessed.[12–15,18,19] Finally, this article represents the outcomes from a single center.

CONCLUSION

This is the first report of a novel surgical approach to TBM: anterior and posterior tracheopexy. Our early surgical outcomes are promising, with most patients reporting improvement in preoperative respiratory symptoms. The RLN is at risk during extensive paratracheal dissection, and true vocal fold immobility was a complication noted in our cohort. The otolaryngologist plays a crucial role in the treatment of TBM, particularly in pre- and postoperative evaluation and assistance.

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