

# Updates on Surgical Repair of Tracheobronchomalacia

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## ABSTRACT

Surgical correction of tracheobronchomalacia (TBM) has continued to evolve over the recent years. A review of the advances in the pre-operative assessment, operative techniques, and post-operative management of patients with TBM was performed. A 3 phase bronchoscopy, as part of the pre-operative evaluation, provides the most information for operative planning for the surgeon. Multiple options for surgical intervention are available, including an open, thoracoscopic, or median sternotomy approach to relieve anterior compression, or an open or thoracoscopic approach for performing a posterior tracheobronchopexy. Excellent outcomes have been seen with both approaches; however, the posterior approach is the preferred method of repair at our institution, as this provides direct support of the posterior membrane and allows for multiple areas of airway collapse to be directly addressed during the same operation; it also makes an anterior approach, if needed, much easier and more effective. Post-operatively, the use of paravertebral catheters has helped tremendously with pain control and respiratory function. The success of TBM repair is due to a multi-disciplinary team approach, as well as experience and analysis of many patients, that can be found at a Center of Excellence due to the high referral rate.

## Introduction

Tracheobronchomalacia (TBM) can be understood as excessive collapse of the airway during exhalation and has a prevalence of approximately 1:2100 births<sup>1,2,3</sup>. Undiagnosed TBM results in persistent airway collapse, which can lead to poor ventilation and poor secretion management resulting in recurrent infections. Over time, this can lead to permanent lung damage in up to 27% of patients with undiagnosed TBM by the age of 8 years old<sup>2</sup>. Two types of tracheobronchomalacia are commonly described in the literature – congenital and secondary. Congenital TBM is thought to be due to abnormal growth with deficiency in the cartilaginous rings; it is usually found with other congenital anomalies, such as esophageal atresia, craniofacial abnormalities, or prematurity<sup>4</sup>. Secondary TBM is degeneration of normal cartilage as a result of extrinsic compression from the cardiopulmonary vasculature<sup>3</sup>, anomalous great vessels, tumors<sup>5</sup>, dilated proximal esophagus, or prolonged intubation/tracheostomy<sup>1</sup>. Risk factors for tracheomalacia include prematurity and esophageal atresia with trachea-esophageal fistula (EA/TEF). In patients with prematurity, the tracheal rings are more compliant, and there may be a lower ratio of cartilage to posterior membrane smooth muscle, predisposing these patients to a more compliant airway<sup>1,5</sup>. In the setting of EA/TEF, not only could there be an anatomical abnormality to the trachea, but the dissection and operative technique of EA/TEF repair could also lead to a weaker and less well-supported posterior membrane<sup>6</sup>. The reported incidence

of EA/TEF in patients with TBM ranges from 11-94%, depending on the study<sup>2,3,7-11</sup>. Congenital heart disease has also been recognized as a common comorbidity and seen in 24-53% of the TBM population<sup>10,11</sup>.

The etiology of symptoms of TBM are related to the intermittent airway collapse, particularly during forceful exhalation, which leads to impaired airway mucus and contaminant clearance and an increased frequency and prolongation of respiratory illnesses<sup>11</sup>. Physicians and parents may be alerted to abnormalities due to patient symptoms: barking cough<sup>9</sup>, dyspnea, gasping, neck extension, increased work of breathing, stridor, exercise intolerance, feeding difficulties<sup>5</sup>, recurrent pneumonia or respiratory infections<sup>7</sup>, blue spells<sup>10</sup>, and even acute life-threatening events (ALTEs)<sup>1</sup>. Now being called BRUEs, or brief resolved unexplained events, these episodes include  $\geq 1$  of the following: cyanosis or pallor; absent, decreased, or irregular breathing; changes in muscle tone; and altered responsiveness that resolve in under 30 seconds – 1 minute with the patient returning to his baseline health<sup>12</sup>. In patients with severe tracheomalacia, ALTEs/BRUEs or blue spells were the most common symptoms in the younger population, while a persistent cough was the most common symptom in older children<sup>9</sup>.

Since the publication of "Direct tracheobronchopexy to correct airway collapse due to severe tracheobronchomalacia: Short-term outcomes in a series of 20 patients", surgical correction of tracheobronchomalacia has continued to evolve rapidly. This article will review the changes and improvements in pre-operative evaluation, operative interventions, and post-operative management.

### Pre-Operative Evaluation

Successful correction of tracheobronchomalacia (TBM) still relies heavily on the pre-operative evaluation for determination of the appropriateness of repair, anatomic anomalies that can be corrected, surgical approach, and technique. The dynamic 3 phase bronchoscopy (rigid or flexible) remains the mainstay, and perhaps the gold standard, in evaluating major and small airway anomalies,<sup>1,3,9,13</sup> including any dynamic posterior collapse of the airway, as well as documenting fixed anterior compression from the great vessels. It is essential to work in conjunction with the anesthesiologists to have the patients lightly anesthetized during the procedure to allow for spontaneous breathing. The ability of the patient to spontaneously breathe and even vigorously cough to stimulation is essential for a proper examination. The initial assessment (Phase 1 of the 3 phase dynamic bronchoscopy) occurs during shallow spontaneous breathing after removal of all secretions. Assessment of vocal cord function is attempted, although any anesthesia can artificially induce unilateral or bilateral vocal cord

dysfunction. The presence of a laryngeal cleft is also evaluated during the initial portion of the procedure<sup>7</sup>. The ultimate goal, however, is to determine if there is any anterior collapse of the airway (fixed stenosis) from the aorta, innominate artery, or aberrant subclavian artery (as can be seen by pulsations of the posterior wall), or other anomalies such as vascular rings, including double aortic arch, etc. Assessment of the shape of the tracheal cartilages is also determined. Wailoo and Emory in 1979 were the first to look at postmortem tracheas in pathology and described the elliptical deformity of the tracheal lumen that is seen in TBM; they subsequently described the C, U, and bow-shaped cartilages that are often seen<sup>3</sup>. With TBM, the anterior cartilaginous to posterior membranous trachea ratio can be reduced from 5:1 down to 3:1 due to the widening of the posterior membrane<sup>7</sup>. This widening, which is seen on bronchoscopy as a change from the normal C-shaped cartilages to U-shaped and then bow-shaped cartilages, allows for more dynamic motion of the posterior membrane, and a higher likelihood that the patient will have significant dynamic airway collapse with forced exhalation, Valsalva, or coughing<sup>3,4</sup>.

The next step (Phase 2) during the bronchoscopic evaluation is to induce coughing or Valsalva maneuvers by the patient. This often challenges the anesthesia team until they gain experience. Airway collapse during this part of the procedure is typically from posterior membrane intrusion. Bairdain described a scoring system that divided the upper airway into five parts<sup>11</sup>. T1 (upper third) is from the cricoid to the thoracic inlet, T2 (middle third) is from the thoracic inlet to the mid-portion of the trachea, and T3 (lower third) is from the mid-portion to the carina<sup>4,11</sup>. The scoring systems also includes the right and left mainstem bronchi. Each area was given a score of 0-5 depending on the amount of airway that was open during both gentle and vigorous breathing (0 -> 90% open, 1: 75-90% open, 2: 50-75%, 3: 25-50%, 4: 10-25%, and 5: <10% open)<sup>11</sup>. T2 and T3 tended to be the areas most affected by TM<sup>2,4</sup>. Severe tracheomalacia, which usually coincides with coaptation (100% airway collapse) in symptomatic patients would be indicative of the need for appropriate effective airway support, often with a posterior tracheopexy and/or bronchopexy.

Phase 3 of the 3 phase dynamic bronchoscopy is then performed with the patient heavily anesthetized or paralyzed; the airways are distended to 40cm H<sub>2</sub>O to look for airway anomalies such as tracheal diverticuli, tracheo-esophageal fistulas, and aberrant bronchi. All these findings are carefully recorded using the airway classification system that was published in the original article. At the time of corrective surgery, it is ideal to repeat the 3 phase dynamic bronchoscopy prior to positioning and draping to confirm previous findings and confirm the plan for intervention.

Another useful tool for pre-operative evaluation is the multi-detector computed tomography (MDCT) scan with a 3D reconstruction of the airway and vascular anatomy, including identification of the Artery of Adamkiewicz<sup>3,11</sup>. The MDCT scan is useful in identifying anomalies of the great vessels, including aberrant arteries, vascular rings, and a circumflex aorta,<sup>1</sup> 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 too far anterior, it will typically push on the mid portion of the left mainstem bronchus (L2), narrowing it, and the posterior intrusion is potentially made worse when the bronchus is brought to the anterior longitudinal spinal ligament during the posterior bronchopexy and tracheopexy. If this is the case, a descending posterior aortopexy is often performed. Identification of the Artery of Adamkiewicz guides the surgeon in avoiding injury to this critical artery during a posterior descending aortopexy. In a study by Ullman et al comparing the sensitivity and specificity of multi-detector computed tomography (MDCT) with bronchoscopy, MDCT was highly sensitive (100%) and had a NPV of 100% and PPV of 90%<sup>13</sup>. Similarly, in a study by Ngercham, TBM was identified on MDCT in 17/18 patients who were also found to have TBM on bronchoscopy. When comparing MDCT to bronchoscopy, MDCT showed excellent specificity and overall accuracy, with a 91% agreement over all tracheal segments<sup>4</sup>. However, patients under 5 year of age were intubated to ensure appropriate inspiratory and expiratory holds for the CT scan; depending on the placement of the endotracheal tube, significant upper tracheomalacia can be missed<sup>14</sup>. Furthermore, the positive pressure that is exerted during patient coughing is hard to artificially reproduce, meaning that MDCT tends to underestimate the degree of airway collapse<sup>3,4</sup>. As a result of some of these downfalls, bronchoscopy and MDCT should be used as adjunctive therapies to determine not only the degree of malacia, but also to search for any vascular anomalies or parenchymal disease prior to recommending surgery<sup>2,7</sup>.

Other testing that can also be considered in the pre-operative evaluation includes pulmonary function tests (PFTs) and an esophagram or barium swallow. PFTs may be very unreliable at diagnosis of TBM, but may give clinicians objective data to assess the degree of improvement after surgery, especially in older children who are able to participate in the testing<sup>3</sup>. In patients with EA, getting an esophagram at the time of the pre-operative workup can help eliminate an esophageal stricture or recurrent fistula as the cause of a patient's feeding problems or cyanotic episodes<sup>6</sup>. There is no role for pre-operative echocardiogram unless the patient had a history structural heart lesions<sup>1</sup>.

A combination of symptom presentation and

bronchoscopic scores used together are best at determining the indications for treatment for the patient once the workup is complete<sup>2</sup>. Mild to moderate symptomatic TBM may improve within a few years without surgical intervention, and these patients can undergo medical management with hypertonic saline to loosen secretions, ipratropium to decrease secretions and stiffen the lower airways, and potentially inhaled steroids if significant inflammation is seen<sup>3</sup>. Pulmonary toilet, chest physiotherapy, and control of gastro-esophageal reflux (GER) is encouraged<sup>3</sup>. In patients with a history of EA/TEF, the concern for GER and need for fundoplication must be seriously considered. In the past, the initial approach for those with severe TBM was placement of a tracheostomy, however this is not a risk-free procedure. Even after the child has undergone successful tracheostomy placement, there are still risks of tracheal injury, inflammation causing granulation tissue, and the formation of a tracheal shelf, tracheoarterial fistula, or tracheal stenosis, tracheal plugging, accidental decannulation, delayed vocalization, and difficulty with decannulation as the placement of the trachea does not address the problem of the collapsible airway<sup>3</sup>. In patients with TBM that extends beyond the tip of the tracheostomy tube, the utility of the tracheostomy is to be questioned since the patient will still require positive airway pressures and may continue to have blue spells and recurrent infections that can cause progression of the TBM. In our experience, long segment TBM that involves distal T3, carina, or the bronchi is an indication for tracheal surgery for correction of the TBM.

Patients who are deeply symptomatic (blue spells, ALTEs, inability to wean on mechanical ventilation<sup>3</sup>, perioral cyanosis<sup>6</sup>) with bronchoscopic findings of severe TBM (categories 4 and 5 on the scoring symptom, or less than 25% opening of the airway), are offered surgical intervention for airway support to the patients/families<sup>3</sup>. The decision of what technique to perform depends heavily on the bronchoscopic findings, results of the MDCT, and associated congenital conditions. Patients found to have coaptation due to significant posterior intrusion, recurrent TEF, or a tracheal diverticulum will be offered a posterior tracheopexy, as the right-sided posterior thoracotomy approach allows the surgeons to address all problems during the same operation. The left-sided approach is typically reserved for the patients with a right aortic arch. For patients who primarily have secondary TBM due to vascular anomalies and innominate artery compression, or who require surgery for a congenital heart lesion, an anterior approach is frequently offered<sup>11</sup>.

## Operative Interventions

Derived from Gross, who in 1948 identified a treatment for innominate artery syndrome and originally described by Filler, anterior aortopexy was the first technique

described to correct tracheomalacia,<sup>3</sup> and it continues to be the most common surgical technique employed. For the anterior approach, either a right or left thoracotomy may be performed, although we strongly prefer a median partial sternotomy<sup>10</sup>. To gain access to the anterior mediastinum via thoracotomy, the pectoralis muscle is elevated, and the chest is entered through the 3<sup>rd</sup> intercostal space<sup>3</sup>. Once inside the chest, a partial or complete thymectomy is performed, and with great attention to the location of the phrenic nerve and recurrent laryngeal nerve, the pericardium is opened anteriorly. Traction sutures are placed on the edges of the pericardiotomy flaps to maintain exposure, and a small subcutaneous pocket is made anterior to the upper sternum for final placement of the tightened sutures<sup>3</sup>. Sutures are passed through the anterior adventitia of the ascending aorta and innominate artery before being attached to the posterior sternum, in either an interrupted or horizontal mattress pattern<sup>10</sup>. Intra-operative bronchoscopy is performed prior to securing the sutures to confirm the relief of vascular compression and opening of the airway. It is important to monitor the blood pressure in the affected extremity (typically the right arm) and the lower extremities to avoid kinking or compressing the innominate artery and aorta with this maneuver. The sutures are tied in the subcutaneous tissue; the thoracotomy incision is then closed. Dr. Kay-Rivest also described a left anterior Chamberlain thoracotomy for aortopexy repair and found that after repair, not only did patients have complete resolution of symptoms, but there was a significantly less readmission rate at 2-year follow-up in aortopexy group<sup>6</sup>.

The challenge in the left anterior thoracotomy approach is to avoid operating on the pulmonary artery, as that structure may be easily confused with the aorta<sup>10</sup>. Inadvertent elevation of the pulmonary artery may potentially lead to kinking and increased impedance across the pulmonary artery, possibly leading to alterations on pulmonary blood flow or right heart failure. We feel it is important to monitor for alterations in blood pressure distal to the areas of blood vessel manipulation because of these maneuvers and perform an echocardiogram before leaving the operating room to ensure that there is no alteration in blood flow or cardiac function<sup>10</sup>.

In 2000, Morabito from the United Kingdom described using an anterior mediastinal approach for 16 patients. The patient is placed in the supine position with a shoulder roll underneath<sup>3</sup>. The operation consisted of a low cervical transverse incision (about 3mm) and then a midline manubrial split to expose the thymus and the underlying ascending aortic arch<sup>8</sup>. The thymus is partially resected, and the pericardium is opened to obtain exposure of the mediastinal structures, including the ascending aorta, innominate artery, pulmonary artery, and trachea<sup>8,10</sup>. This

approach allows for better exposure of both the great vessels as well as the trachea and bilateral mainstem bronchi<sup>10</sup>. Pledged sutures are placed on the ascending aorta and the innominate artery with careful consideration to not disrupt the pre-tracheal fascia; it is this fascia that will indirectly lift the anterior trachea when the aortopexy sutures are tightened<sup>14</sup>. Intra-operative bronchoscopy is performed to ensure adequate opening of the trachea<sup>8</sup>. If compression of the airway still persists, especially in the left mainstem bronchus, sutures can be placed in the pulmonary artery for anterior fixation. In very severe cases, sutures are placed directly through the tracheal cartilages and are secured to the sternum to ensure opening of the trachea<sup>3,10</sup>. Instead of placing sutures directly through the aortic adventitia, some surgeons prefer to leave the pericardium intact and use the pericardium for pexy sutures to indirectly open the trachea<sup>10</sup>. Approaching anterior tracheal compression from a sternotomy, as compared to the lateral thoracotomy approach allows for better access should bleeding occur, and helps to ensure that both sides of the trachea are elevated equally<sup>10</sup>. This approach has less morbidity than an anterior thoracotomy and a more appealing cosmetic appearance<sup>8</sup>.

Most recently, the minimally invasive thoracoscopic approach has been used to relieve anterior vascular compression. In these incidences, the patient is placed supine with either a left or right-sided tilt, 3 ports (1 camera and two working ports) are placed, and the chest is insufflated with 5mmHg of CO<sub>2</sub>; the phrenic nerve is identified, the thymus is dissected free, and the pericardium is opened<sup>10</sup>. The right side is generally preferred, as using a left-sided approach may be confusing and lead to pulmonary artery suspension instead of aortic suspension. 3-0 or 4-0 Ethibond sutures are passed through the sternum anteriorly, transversely through the ascending aorta or pericardium, and then back through the sternum to be secured in the subcutaneous tissue with use of a Reverdin needle<sup>5,15</sup>. Bronchoscopy is performed both at the beginning, during the suture tensioning as testing, and at the end of the case to determine that the airway will remain open. This technique, with operator modifications, has been reported by us<sup>10</sup> and also performed in the Netherlands by Dr. van der Zee, et al.<sup>16</sup> In studies done by both Dr. Jennings and Dr. van der Zee, significant improvement in symptoms and ability to wean off respiratory support was achieved; however, a small subset of patients required future intervention due to recurrence of symptoms<sup>5,16</sup>. This procedure was also shown to be safe in young children, with the youngest patient in Perger's study being 6 weeks of age<sup>5</sup>. In a retrospective review of our patient cohort comparing lateral thoracotomy, thoracoscopy, and partial median sternotomy for the relief of anterior compression and tracheomalacia, we found superior results with the

use of a partial median sternotomy compared to the other approaches<sup>10</sup>. The advantages of a partial median sternotomy include the ability to use pledgeted sutures, ability to relieve compression in many areas, such as the innominate artery, aorta, and pulmonary artery, and ability to provide direct support of the trachea and bronchi. This has led us to prefer the partial median sternotomy approach for anterior tracheal work<sup>10</sup>.

For patients with very proximal TBM, a cervical approach may be employed. In a case report by Bjornson, a child presented with high (cervical) TM as well as a proximal missed TEF. Using a similar incision approach as the partial sternotomy, a collar incision was used, the thyroid was mobilized, and the tracheoesophageal groove was identified to dissect the esophagus and divide the TEF. Dissection was then carried underneath the manubrium, and the anterior surface of the trachea was secured to the posterior manubrium using interrupted absorbable polydioxanone sutures<sup>14</sup>. In patients undergoing a sternotomy for anterior work, the dissection and repair can be extended to the cervical trachea through the same incision, if required.

Currently, our practice has morphed to performing a right posterior thoracotomy (in those children with a left sided aortic arch) and a posterior tracheobronchopexy with intra-operative flexible bronchoscopy<sup>2</sup>. From this approach, direct support of the posterior membrane and relief of the posterior compression of the trachea and both mainstem bronchi can be achieved in one intervention. In the posterior approach, an incision is made in the 4<sup>th</sup> intercostal space, and a muscle sparing technique is used to dissect down to the ribs<sup>2</sup>. Once inside the chest, the esophagus, back wall of the trachea, and aorta are dissected<sup>3</sup>. The left vagus nerve, thoracic duct, and left recurrent laryngeal nerve are identified and preserved. If the patient also has a recurrent TEF or a tracheal diverticulum, it is resected at this time. If the descending aorta is located anterior to the spine and causing posterior compression of the left mainstem bronchus, autologous pledgeted polypropylene sutures are placed in the adventitia of the descending aorta and secured to the left side of the spine prior to addressing the trachea<sup>3</sup>. Both upper and lower extremity blood pressures are monitored carefully, preferably with arterial lines, during securing of the sutures to ensure distal aortic flow is not obstructed. Under bronchoscopic guidance, sutures then are placed into the posterior tracheal and mainstem bronchial membrane in a horizontal mattress pattern before being secured to the anterior longitudinal spinal ligament<sup>2</sup>. Continuous bronchoscopy is performed to ensure that the sutures are not entering the tracheal lumen during placement. Tension is placed on all the tracheal sutures to ensure adequate opening of the airway without distortion prior to tightening of the sutures<sup>2</sup>. At

our institution, our anesthesia colleagues will perform a negative suction test (the Munoz Maneuver, named after Dr. Carlos Munoz). While looking through the bronchoscope, they will attach suction tubing to the endotracheal tube and slowly increase the negative pressure while looking at the airway with the flexible bronchoscope; the pressure is increased to -40 or -50cm of water. If the airway remains open, then no further intervention is required; if the airway is inappropriately disfigured or still collapsing, then further work to relieve the obstruction is undertaken. We have also changed to using only autologous pledgets for our tracheopexy sutures (from native pleura or ligated and resected azygous vein) as there is a risk of developing erosion into the esophagus by felt pledgets next to the esophagus.

In a recent study by Shieh, the largest series to date reporting on outcomes following posterior tracheopexy, significant reduction of symptoms, such as cough, noisy breathing, recurrent respiratory infections, oxygen dependence, and ventilator dependence after posterior tracheopexy was seen.<sup>2</sup> Most importantly, there was no recurrence of ALTE/BRUE events following the posterior tracheopexy.<sup>2</sup> TBM scores also significantly improved in all 5 regions of the large airways<sup>2</sup>. Likewise, in Bairdain's work, there was significantly less TBM in all regions of the trachea on the follow-up bronchoscopy using the scoring system, with the greatest areas of improvement being in T2 and T3<sup>11</sup>. There was no recurrence of ALTEs, although one did require a repeat operation. There was also significantly less documented incidence of pneumonia and need for supplemental oxygen, although this was not eradicated completely<sup>11</sup>. Even in patients with some degree of anterior intrusion, most symptoms will resolve without need for further intervention following posterior tracheobronchopexy<sup>3</sup>. Anterior work (aortopexy and anterior tracheobronchopexy) is considered as a second procedure if the patient continues to have persistent symptoms. These cases are uncommon, approximately 15%, and are typically due to either bow shaped cartilages with a descent of the anterior tracheal cartilages over the anterior spine (mimicking posterior membrane intrusion), or those with more than 50% anterior tracheal intrusion from vascular compression. Rarely at our institution is the anterior support work performed first as the anterior translation of the trachea may make the persistent posterior intrusion difficult to treat. However, in cases where there appears to be isolated severe anterior vascular compression of the trachea or structural congenital heart disease that needs surgical repair, we may proceed with anterior tracheal support first. In the few cases where this has been inadequate, we have been able to do the posterior work later with success.

In certain patients (such as older children or teenagers)

with tracheobronchomalacia, thoracoscopic, or even robotic repair is being introduced into the surgeon's armamentarium. Through a right-sided approach, the esophagus is mobilized, and the posterior tracheal membrane is sutured to the anterior longitudinal spinal ligament<sup>15</sup>. In Jennings' study comparing approaches for aortopexy, there was a 38% recurrence rate in the thoracoscopic group requiring repeat surgery;<sup>10</sup> Dr. van der Zee also reported a 31% recurrence rate with the thoracoscopic approach, potentially indicating the learning curve that is needed whenever a new technique is introduced.<sup>16</sup> With an open aortopexy, there is approximately a 10% reported incidence of persistent symptoms requiring a further operation<sup>2</sup>.

Around a 15% complication rate has been reported following surgery for TBM,<sup>3</sup> including pericardial effusion for those patients in which the pericardium was opened, bleeding, vocal cord paralysis<sup>8</sup>, phrenic nerve injury (with 2 documented patients requiring diaphragmatic plication), chylothorax requiring prolonged chest tube and diet modification<sup>3</sup>, or narrowing of the esophagus at the level of the thoracic inlet in patients who undergo a high posterior tracheopexy that may require endoscopic dilation.

One of the limitations in performing either a posterior tracheopexy or anterior aortopexy is found in the subgroup of patients that also have significant small airway disease, most often due to bronchopulmonary dysplasia (BPD). Surgical intervention may be beneficial if there is significant large airway collapse; however, some patients will still require supplemental oxygen or mechanical support due to the persistence of their small airway and alveolar disease. It is important to be up front with families about the capabilities, but also the limitations, of surgical intervention in patients with multiple level airway problems.

Other options reported include splinting and stenting of the airway. Splints can be created either from autologous rib cartilage, mesh, or ceramic rings; Green et al. have reported trialing the use of custom-made 3D printed external splints for the treatment of left mainstem collapse<sup>17</sup>. However, the long-term effects, including compression of the trachea during the growth of the child and potential for erosion, are unknown<sup>3</sup>. Stent placement has the appeal as this procedure is less invasive and has a shorter recovery times for the patient. However, with the high risk of clogging or migration (up to 43%)<sup>3</sup>, it was found that stenting had higher failure rates and more severe morbidity and mortality than aortopexy and has subsequently been less popular unless specific situations arise<sup>9,3</sup>.

### Post-Operative Management

The advancement in post-operative management has also progressed rapidly. The use of a paravertebral catheter pain regimen has decreased the need for post-

operative pain medication and has allowed for better respiratory recovery<sup>18</sup>. A retrospective study looking at the use of paravertebral catheters in patients with long gap esophageal atresia who undergo thoracotomy during repair showed decreased opioid and benzodiazepine use as well as decreased intensive care unit (ICU) length of stay<sup>18</sup>; we believe that these same results also apply to our tracheomalacia patients. Unless a patient was on a ventilator prior to surgery, an attempt at extubation is performed in the operating room prior to transfer to the ICU. This has led to a decrease in both ICU length of stay and overall hospital stay in patients requiring only TBM repair. In the past two years, the median length of stay in the ICU for these patients has been 2 days at our institution, with the biggest contributor to ICU stays being pain control with subsequent breath holding episodes. The median length of hospital stay for patients undergoing isolated airway work (excluding any esophageal intervention) in the prior 2 years was 7 days, with an average of 12.6 days, which has decreased dramatically from the 26 days that we reported prior. Due to the success of the posterior tracheopexy in symptom relief and degree of post-operative airway collapse<sup>2</sup>, we do not routinely perform a bronchoscopy during the immediate post-operative period unless the patient is symptomatic. Long-term follow-up at our institution includes a surveillance rigid or flexible dynamic 3 phase bronchoscopy at 1 year out from the time of operation unless the patient has symptoms.

A literature review done by Torre in 2012 showed that out of 758 patients who had aortopexy, 80% improved significantly at a median follow-up of 47 months<sup>9</sup>. Similarly, Morabito looked at 16 patients after anterior aortopexy from a partial sternotomy approach; 10/16 did not require any post-operative mechanical ventilation, and 14/16 did not require any past the first 24 hours. One patient required CPAP for 4 months, and one patient required a tracheostomy due to bilateral vocal cord paresis<sup>8</sup>. In Jennings' study, all groups showed symptom improvement and decreased work of breathing with 2/41 patients still requiring ventilatory support<sup>10</sup>.

In an initial study looking at the different surgical approaches for aortopexy, Jennings et al. noted that 4/5 patients who had a tracheostomy placed for TBM were able to be decannulated<sup>10</sup>. We have since been able to expand upon that, including patients that had both anterior and posterior aortopexies. In a subset analysis of patients with a tracheostomy in place due to tracheobronchomalacia, 20/25 were either decannulated, ready to begin the decannulation process, or were on trach collar but required further laryngeal surgery due to subglottic stenosis that prevented full decannulation, confirming the potential benefits of surgical intervention for tracheobronchomalacia<sup>2,11,19</sup> (unpublished).

## Conclusion

The treatment of tracheobronchomalacia continues to evolve, with many options available depending on the type of airway seen. A thorough pre-operative workup, including bronchoscopy and MDCT are crucial for intra-operative planning and family counseling. Results have shown significant symptom improvement, especially with posterior tracheopexy in appropriate cases. However, as standardization and long-term outcomes are still in evolution, specialized care at Centers of Excellence can only enhance the understanding and effectiveness of treatment of these complex airway problems.

## List Of Abbreviations

TBM – tracheobronchomalacia

CT – computed tomography

ALTE – acute life-threatening events

BPD – bronchopulmonary dysplasia

ICU – intensive care unit

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