Surgical approaches to aortopexy for severe tracheomalacia

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Abstract

Purpose: The purpose of this study was to determine the outcomes among three different surgical approaches for performing an aortopexy to treat severe tracheomalacia (STM).

Methods: A retrospective review was performed for all patients who underwent an aortopexy by pediatric surgeons at a single institution during 1997–2012. Data collected included details of the operative approaches and clinical results. The data were analyzed using Chi-square and Fisher exact test.

Results: Forty-one patients underwent an aortopexy. The operation was chosen by the surgeon and not randomized. Exposure was by partial sternotomy (PS) (20), open thoracotomy (12), or thoracoscopic approach (7). Only the PS approach was done by a single team. All groups showed improvement in work of breathing, prevention of severe respiratory distress, and acute life threatening events. These effects were more dramatic for the PS group, especially regarding oxygen and/or ventilator dependence and the ability to undergo tracheostomy decannulation. Among the sixteen patients with failure-to-thrive before successful aortopexy by any technique, ten demonstrated significant improvement in their growth (p = 0.025). The recurrence rate for the thoracoscopic approach was 38%, and there were no recurrences in the partial sternotomy and the thoracotomy groups, 38% vs 0% vs 0%, p = 0.005. Simultaneous bronchoscopy was utilized more commonly in the PS group compared to the thoracotomy and thoracoscopic group, 95% vs 62% vs 38%.

Conclusions: In this series, the partial sternotomy technique had the most reliable resolution of symptoms and no recurrence requiring reoperation. The PS approach to STM has the technical advantages of an improved exposure with equal access to the vessels over the right and left mainstem bronchi, as well as the trachea and a more specific elevation of the arteries, including suspension of the pulmonary arteries and trachea itself when desirable. Simultaneous bronchoscopy during aortopexy and an experienced team also likely contribute to improved outcomes. The variations in populations, follow-up, and use of continuous intraoperative bronchoscopy, however, make firm conclusions difficult.

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Severe tracheomalacia (STM) and bronchomalacia may require treatment which is most commonly accomplished by an “aortopexy”. In this paper, aortopexy is the operation, but the procedure details may involve suture support of other mediastinal structures in addition to the aorta. By pulling the overlying vessels and other structures anteriorly to open up the airway, an aortopexy limits collapse during the active exhalation phase of breathing; however, it does not directly treat tracheomalacia. With the airway attached anteriorly to the vessels and posteriorly to mediastinal structures by connective tissue, suture elevation of the aorta and, usually, the innominate artery to the back of the sternum will maintain an adequate airway. By this indirect mechanism, aortopexy has been found to be generally beneficial in relieving the consequences of STM.

An effective aortopexy, we believe, has several components. Foremost is bringing the vessels up nearly to the back of the sternum which elevates the anterior aspect of the trachea and may, also open the proximal portion of the main stem bronchi. Occasionally, with significant malacia of the mainstem bronchi, elevation of the pulmonary arteries and even the adjacent pericardium must be carried out to hold the mainstem bronchi open. Occasionally these maneuvers have proven insufficient, and direct suture placement into the tracheal and/or bronchial cartilage has been required to provide adequate airway support. The sutures placed in the vessel walls are of obvious importance and must balance holding power without causing bleeding. Finally, because an aortopexy is an indirect solution, the ability to assess the effects of the elevation of the vessels and other structures on the airway by bronchoscopy while the procedure is being carried out is valuable to ensure the desired result.

Several surgical approaches have been used to reach the arteries and bring them forward to the sternum. Right and left anterior as well
as more lateral thoracotomy incisions have been used and also cervical, mediastinal and thoracoscopic approaches have been reported [1–3]. The many techniques indicate that, so far, no one approach has proven superior for all cases.

We report a review of all patients who underwent aortopexy by one of three techniques by pediatric surgeons over 15 years at a single institution. This review allowed comparison of the standard open thoracotomy and thoracoscopic techniques with the more recently used partial sternotomy (PS) approach.

1. Materials and methods

A retrospective chart review was performed on all patients who underwent an aortopexy for STM by pediatric surgeons between 1997 and 2012 at Boston Children’s Hospital (IRB P00007671). The patients were grouped by the operative approach and the preoperative status, and the aortopexy results were reviewed.

The operative approach was chosen by the surgeon and not randomized. Only the PS procedures were done by a single surgical team. The patients in this review often had a complex clinical history and frequently STM was not the only significant airway or pulmonary problem. Among the partial sternotomy patients for example, 17/20 (85%) had a prior EA repair done elsewhere and many had significant pre-transfer complications, further compromising overall pulmonary function.

Among the clinical manifestations evaluated as ascribable to STM were: acute life-threatening events (ALTE) defined as a significant apneic episode, or a “blue spell” or transient breathing distress with marked air hunger and/or very loud breathing. Oxygen dependence was less specific but was considered present when supplemental oxygen was necessary to maintain a saturation of 90%. Similarly, it was judged whether or not tracheomalacia was mainly responsible for preoperative intubation or for the presence of a tracheostomy. All patients were evaluated for other causes of respiratory compromise including laryngeal and upper airway difficulties and previous correction of trachea-esophageal fistulas with large residual tracheal pouches producing significant airway intrusion. All patients were evaluated for gastro-esophageal reflux which was treated as appropriate with fundoplication and/or medications.

Evaluation of TM was performed by rigid bronchoscopy while spontaneously breathing in all patients, and more recently also by dynamic airway CT. The TM was classified by location in the trachea (upper third, middle third, lower third) and mainstem bronchi (right, left), and by the degree of airway collapse during exhalation. The TM was judged to be severe when there was near complete collapse of the airway or coaptation of the tracheal walls shown either by bronchoscopy and/or or CT.

The operative data tabulated included the method of aortopexy performed, the details of the elevation of the vessels and pericardium and the use of intraoperative bronchoscopy. Postoperative data included symptom resolution, evidence for recurrence of TM, or the need for reoperation.

Operative Methods: Partial sternotomy: A 3-cm transverse incision was made at the manubrial–sternal junction and a partial upper sternotomy along with partial thymectomy allowed the upper portion of the pericardium to be opened to reveal the innominate artery and ascending aorta. The vessels well-visualized, partial thickness horizontal mattress sutures of pledged 5-0 Tevdek were placed into the antero-lateral aspects of the artery walls (Fig. 1). A flexible bronchoscope passed down the endotracheal tube assessed the result. When the mainstem bronchi remained collapsed despite innominate artery and aortic elevation, sutures were placed in the wall of the pulmonary artery and, for some, in the pericardium adjacent to the pulmonary artery. In a very few cases, non-pledged sutures were also placed in the tracheal and/or bronchial cartilages for more direct elevation of the trachea and bronchus. The sutures were then placed into the posterior sternal fascia or through the sternum itself. A final evaluation of the airway was made by bronchoscopy and an echocardiogram assessed the great vessels after closing.

Thoracoscopic aortopexy (Illustration 2): The thoracoscopic approach has previously been described [4,5]. Three left-sided ports were typically used, although a right sided approach was also done. The sutures were placed along the most anterior portion of the upper ascending aorta and into the sternum (Fig. 2). In 3 cases the sutures were placed into the pericardium overriding the aorta without opening the pericardium. Bronchoscopy was sometimes performed during the case, and was usually done at the end of the case to assess the aortopexy effects on the airway.

Thoracotomy: A left or right anterior thoracotomy incision allowed the pericardium overriding the aorta to be opened. Sutures were passed through the anterior aspect of the upper ascending aorta and

![Fig. 1. Partial sternotomy technique. After performing a partial sternotomy, removing a portion of the thymus and opening the upper portion of the pericardium for exposure, the innominate artery and aortic arch are elevated and sutured to the sternum. This exposure allows pledgeted horizontal mattress to be placed in the lateral aspect of the arteries producing a more even elevation and also the ability to continuously observe the effect by bronchoscopy.](image1)

![Fig. 2. Thoracotomy technique. After performing a limited anterior thoracotomy, either right or left, the near thymic lobe is removed and the ascending aorta and sometimes the innominate artery is sutured to the sternum. These sutures are placed along the anterior surface of the arteries.](image2)
occasionally into the innominate artery and then sutured into the posterior sternum (Fig. 3). Bronchoscopy assessed the result in some during the operation and in most at the end. The operative groups were not strictly comparable although a single surgical team performed the PS group. With these limitations, the data were analyzed using Chi-square and Fisher exact test and the p-values calculated. Statistical significance was considered as p < 0.05.

2. Results

The demographic data are presented in Table 1. The male/female ratio was 20/21 (49%/51%). The large number of EA patients in the PS group reflects the recent development of an Esophageal Atresia Treatment Program with general association of TM in EA patients. The VACTERL association was present in 18/41 (44%) and significant congenital heart disease in 10/41 (24%). The operative details are presented in Table 2. PS utilized intraoperative bronchoscopy more frequently and was the only technique which exposed both the innominate and the pulmonary arteries for suspension. The innominate artery most impinges on the mid-lower tracheal producing the symptoms of TM and its elevation directly rounded up the trachea. Post-operatively, many patients required several days of intubation to clear bronchorrhea.

The before and after comparisons of the methods of aortopexy on the consequences of STM are presented in Table 3. Length of follow-up for all patients was between 10 months and 14 years. Partial sternotomy and thoracotomy had the highest rate of resolution of ALTE symptoms after surgery, while three (37.5%) of thoracoscopic patients had return of symptoms. Many patients required positive pressure support before surgery and the majority of them were weaned off positive pressure support in the immediate post-operative period. Similarly, most patients who required oxygen to maintain oxygen saturation >90% were weaned off oxygen after surgery. The only two patients who required ventilation support after surgery were in the thoracotomy group, one eventually died (Table 4).

There were five patients who had tracheostomies placed for severe TM prior to partial sternotomy, and all but one was subsequently decannulated. Forty-five percent of patients in the PS and thoracotomy groups had failure to thrive (defined as less than the 3rd percentile in weight for age), and 10/15 significantly improved after aortopexy.

PS—Airway symptoms resolved in all patients and all except one patient with tracheostomies had them removed. One patient had ongoing aspiration from cord paralysis. No patients required reoperation for recurrent TM.

Table 1

<table>
<thead>
<tr>
<th>Operative approaches</th>
<th>Partial sternotomy</th>
<th>Thoracotomy</th>
<th>Thoracoscopy</th>
<th>Total</th>
</tr>
</thead>
<tbody>
<tr>
<td>N (%)</td>
<td>20 (49 %)</td>
<td>13 (32%)</td>
<td>8 (19%)</td>
<td>41 (100%)</td>
</tr>
<tr>
<td>Age at aortopexy (months), median (range)</td>
<td>8.6 (3–134)</td>
<td>4.6 (1–36)</td>
<td>4 (1.5–24)</td>
<td>7.5 (1–136)</td>
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<tr>
<td>Esophageal atresia</td>
<td>19 (95%)</td>
<td>8 (62%)</td>
<td>7 (88%)</td>
<td>34 (83%)</td>
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<tr>
<td>-Long gap (&gt;3 cm)</td>
<td>8 (40%)</td>
<td>1 (8%)</td>
<td>2 (25%)</td>
<td>11 (27%)</td>
</tr>
<tr>
<td>-With TEF</td>
<td>15% (73%)</td>
<td>7 (54%)</td>
<td>5 (63%)</td>
<td>27 (66%)</td>
</tr>
<tr>
<td>-EA repair done elsewhere</td>
<td>17% (85%)</td>
<td>4 (31%)</td>
<td>0 (0%)</td>
<td>21 (51%)</td>
</tr>
<tr>
<td>Associated bronchomalacia a</td>
<td>13% (65%)</td>
<td>6 (46%)</td>
<td>3 (38%)</td>
<td>22 (54%)</td>
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<tr>
<td>Previous aortopexy</td>
<td>2% (10%)</td>
<td>1 (8%)</td>
<td>0 (0%)</td>
<td>3 (7%)</td>
</tr>
</tbody>
</table>

a Bronchomalacia defined as collapse during expiration on bronchoscopy and/or airway CT.

b Prior aortopexy elsewhere during correction of tetralogy of Fallot.

c One patient also had a proximal TEF.
3. Discussion

Table 3

<table>
<thead>
<tr>
<th>Operative approaches</th>
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<th>Thoracoscopy</th>
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<tbody>
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<td>N (%)</td>
<td>20 (49%)</td>
<td>13 (32%)</td>
<td>8 (19%)</td>
<td>41 (100%)</td>
</tr>
<tr>
<td>Aortopexy</td>
<td>20 (100%)</td>
<td>13 (100%)</td>
<td>7 (88%)</td>
<td>40 (98%)</td>
</tr>
<tr>
<td>Innominate artery</td>
<td>20 (100%)</td>
<td>5 (38%)</td>
<td>2 (25%)</td>
<td>27 (66%)</td>
</tr>
<tr>
<td>Pulmonary artery</td>
<td>3 (15%)</td>
<td>0 (0%)</td>
<td>0 (0%)</td>
<td>3 (7%)</td>
</tr>
<tr>
<td>Suspension</td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pericardiopexy</td>
<td>7 (35%)</td>
<td>0 (0%)</td>
<td>3 (38%)</td>
<td>10 (24%)</td>
</tr>
<tr>
<td>Simultaneous bronchoscopy</td>
<td>19 (95%)</td>
<td>8 (62%)</td>
<td>3 (38%)</td>
<td>30 (73%)</td>
</tr>
</tbody>
</table>

Open thoracotomy—No patients required reoperation for recurrence of symptoms but symptoms persisted in three of the 13 patients.

Thoracotomy—Of the eight patients who had thorascoscopic aortopexy, three required subsequent reoperations for recurrence of symptoms. Of the thorascoscopic recurrence patients, one did not have intra-operative bronchoscopy and had a revision aortopexy 1 month later. A second patient had intra-operative bronchoscopy but had a pericardiopexy instead of an aortopexy. Symptom recurrence required re-operation by thoracotomy 9 months later. The third patient had intra-operative bronchoscopy during the thorascoscopic aortopexy, but the symptoms recurred and required re-operation by thoracotomy 1 month later.

3. Discussion

Tracheomalacia occurs with a number of conditions including esophageal atresia, which was the predominant association in this series. The consequences of STM comprise a wide spectrum ranging from breathing and feeding difficulties, a barking or seal-like cough, expiratory stridor, to apneic (blue) spells and ventilatory arrest which can lead to sudden death. STM is especially significant because with increasing ventilatory effort the airway collapse becomes steadily greater and may result in rapidly worsening symptoms. For these patients STM often deserves surgical treatment.

Spontaneously breathing patients who have nearly complete collapse and coaptation of the anterior and posterior walls on exhalation can be said to have severe structural tracheomalacia [6-12]. Our decisions to proceed with surgical intervention in asymptomatic children with STM met this anatomic requirement as confirmed by bronchoscopy and often dynamic airway CT in the PS patients.

In this report, we reviewed a series of patients who had undergone one of several types of aortopexy. All approaches showed improvement in the symptoms of TM after aortopexy, although the benefits were greater in the PS group. The durability of the methods appeared to vary, with open thoracotomy and PS having more lasting results perhaps because of the ease of using of pledged suture and the ability to place more sutures, elevate more structures, or even directly support the trachea. Thoracotomy had less durability with three of eight patients requiring reoperation for recurrence of STM symptoms.

The PS approach appears to offer several advantages. With better exposure, it allows direct visualization of the innominate artery and aorta and makes possible suspension of the right pulmonary artery, the main pulmonary artery as well as the pretracheal fascia and even the trachea. The latter is particularly important in cases where posterior membranous intrusion into the airway increases the effect of STM.

With the direct visualization of the vessels, more accurate placement and directional pull of the sutures can be made with more effective elevation. We have found that the innominate artery which is the principle offender in producing the consequences of severe TM is directly accessible and elevation contributes the most to the benefit of an aortopexy. In the supine position, bronchoscopy is also more accurate and unrecognized distortion of a mainstem bronchus is unlikely to happen. Finally, if bleeding occurs, the direct access will make it easier to control.

One of the difficulties with lateral approaches to aortopexy by thoracotomy and thoracotomy is that the surgeon approaches the midline structures from one side, which means the elevating sutures will favor that side. It is also more difficult to get to the opposite side of the mediastinum for directional pulling on the aorta, or for elevating the opposite side pulmonary artery, pericardium, and trachea or bronchi if needed.

The advantage of the PS approach is direct access to all the mediastinal structures, and elevation of both the trachea and mainstem bronchi can be achieved as needed. In effect, the elevation will be level with no tilt to one side or the other. PS is also amenable to continuous intra-operative bronchoscopy allowing the effectiveness of the aortopexy sutures to be determined. If elevation of the innominate artery and the aorta does not provide adequate tracheal and bronchial opening, then additional traction sutures can be placed in the pulmonary arteries which are often effective for bronchomalacia. Additional sutures placed in the pericardium, or even in the trachea and/or bronchial cartilages directly have on occasion been required for effective treatment. Our data also support the use of intra-operative bronchoscopy and completion echocardiograms as useful adjuncts to successful aortopexy.

The weaknesses of the retrospective review are the variation in patient populations, ages at operation, in surgical teams, use of continuous intraoperative bronchoscopy to assess results, and length of follow-up. These variations limit the value of comparing these techniques.

4. Conclusion

This retrospective review of three non-randomized methods of aortopexy suggests that the apparent technical advantages of the PS
approach will result in a more effective treatment of severe broncho-tracheomalacia with good durability over the short-term. PS is now our preferred approach for operative treatment of these patients.

### References


### Discussion

### "Aortopexy for Severe Tracheomalacia" Presented by Russell Jennings, Boston, MA.

**Unidentified speaker (moderator):** Rusty, can you talk to us about the bronchoscopy? Do you use a rigid scope that also delivers the anesthesia or a flexible scope, through your tube?

**Response: Russell Jennings:** Great question. All of these patients are evaluated preoperatively with rigid bronchoscopy in a spontaneously ventilating patient. You need the dynamics to understand the problem. During the operation we use a flexible bronchoscope through the endotracheal tube and have it set up on the monitor so that the anesthesiologist and the surgeons can see it simultaneously. It is continuously done, and when the child gets a little hypercarbic, particularly in a small child, we’ll remove it for a little while and then we put it back in. Then at the very end it is either a flexible or a rigid bronchoscope with the child spontaneously breathing if possible.

**Discussant: Dr. Arnold Coran (Ann Arbor, MI):** Rusty, nice presentation of a very large series. I’m curious what the indications for aortopexy were in the patients that were not having severe cyanotic spells and sudden infant death-type symptoms, because in this day and age I think we tend to be doing fewer aortopexies and only doing it in that group. My second question is, in the thoracotomy group and in the thorascopy group, did you dissect off the left lobe of the thymus to get a good clear exposure of that ascending aorta (and/in) the pericardial reflection? That technique of course was described by Dr. Gross in his 1953 book. Being at the same institution I am wondering if you followed that same type of operative approach.

**Response: Russell Jennings:** We do believe in our history. In regard to acute life-threatening events, not all the patients have acute life-threatening events. Some of them had recurrent respiratory infections, sort of continuous respiratory infections and some of them have pretty severe respiratory compromise without life-threatening events, so I think we can loosen up the criteria for performing aortopexy. You don’t have to actually die to get one or try to die but you can have recurrent respiratory infections and severe compromise. None of these patients had mild problems.

As far as the thymus is concerned, a variety of surgeons have done these procedures, so there is some variability. From the thorascoscopic approach, most of them have just been mobilizing the thymus, pushing it out of the way and bringing the aorta forward. For the thoracotomy, we typically will remove the thymus on the side that we’re operating on and for the median sternotomy we usually take out one lobe of the thymus.

**Discussant: Dr. Robert Touloukian (New Haven, CT):** Rusty, perhaps you could comment on the genesis of the tracheomalacia insofar as the development of the esophagus is concerned. Many of these patients have a partially obstructed esophagus, either inborn as a result of their esophageal atresia or secondary to the operation itself with stricture or reflux. Perhaps you could comment on any additional esophageal procedures that might be necessary or the investigation required before you proceed with aortopexy.

**Response: Russell Jennings:** That is an extremely complicated question. Tracheomalacia is usually revealed by the aorta leaning on a little segment of the trachea. For the esophageal atresia patients it seems there is more than one type and sometimes we’ll get the upper part of the trachea compressed and perhaps that is because of the in utero large pouch, and is more of true intrinsic malacia of
the trachea. Sometimes we'll see long segment posterior intrusion so there is a lot of excess posterior membrane and that becomes a different problem.

Robert Touloukian: What about the secondary operations on the esophagus before you proceed with aortopexy?

Russell Jennings: Yes, so my current approach to tracheomalacia is that we try to get all the posterior trachea work done, do all the esophageal work, get that done, and I think often and I would say even the majority of the time, in kids who have pretty significant tracheomalacia, if the esophagus is fixed and you can move it a little bit off to the side, they seem to have a marked improvement in the symptoms of tracheomalacia. Those kids who don't are the ones who have the large tracheal pouch left behind and sometimes that becomes quite floppy and can intrude into the airway along the whole length of the distal half or third of the trachea. For those kids we have started doing a posterior tracheopexy at the time of the esophageal atresia repair, so we can just tack that down and that seems to be helping but we haven't done enough of those yet to be able to report it.

Discussant: Dr. David Sigalet (Calgary, Alberta): I was interested in the comment about tacking the pulmonary artery. Do you bring it all the way forward and oppose it to the sternum or do you leave little guide wire stitches? I've seen that done before. I've done it myself. Every time I do it I'm very nervous.

Response: Russell Jennings: Yes, yes to both. If it's the main pulmonary artery it's pretty easy to bring all the way forward if necessary. That's the beauty of this operation. If you bring everything too far forward, particularly the pulmonary arteries, you'll take the trachea from being flat in an AP dimension to being narrow in the lateral dimension, so you have to bring it up sort of just right. We find that sometimes we bring up the innominate artery and left side of the aorta and the main pulmonary artery and that's all we want to do. Sometimes we will leave a little bit of Silastic behind because on the redo aortopexies it's really hard to get in when it's totally fused to the posterior sternum.

Unidentified speaker: It seems like that's where bronchoscopy really helps you out because you can see — I mean I can't stand doing these without taking a look.

Response: Russell Jennings: Absolutely essential to have continuous bronchoscopy and I would advocate for an echocardiogram at the end to assess the effects on the arteries.

Unidentified speaker: To what do you attribute the thoracoscopic failures and do you now recommend against that approach?

Response: Russell Jennings: No, I'm not totally against it. I think the thoracoscopic approach has always been a few simple sutures going through the aortic wall up to the sternum and lacks precision. I think the addition of pledgets might make them more durable.