Operative Techniques in Thoracic and Cardiovascular Surgery

Thoracoscopic Aortopexy for the Treatment of Pediatric Tracheomalacia: A Right-Sided Approach

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Tracheomalacia (TM) refers to a softening of the cartilaginous tracheal rings. The weakened cartilage allows a narrowing in the anteroposterior tracheal diameter. Its incidence is estimated to be 1 in 1445 infants. Both congenital and acquired forms have been identified. Congenital TM is most commonly associated with tracheoesophageal fistula/esophageal atresia in children.[1] The effected tracheal rings have been found to contain a shortened segment of cartilage with a compensatory lengthening of the membranous portion.[2] This is thought to be caused by abnormal embryonic separation of the trachea from the esophagus, leaving the trachea with too much tissue.[1] Primary TM has also been associated with immature cartilage in premature infants, primary cartilage disorders such as polychondritis and chondromalacia, and several syndromes including the mucopolysaccharidoses and Down syndrome. The acquired or secondary forms of TM are more common than the primary forms and result from prolonged endotracheal intubation or tracheostomy. Secondary TM can also be the result of external compression from surrounding cardiovascular anomalies, skeletal disorders such as scoliosis, and space-occupying tumors or cysts.[3]

The weakened trachea is subsequently susceptible to collapse with increased intrathoracic pressure, as in expiration, coughing, and crying. It is also vulnerable to external pressures created by food boluses, dilations proximal to esophageal strictures following tracheoesophageal fistula/esophageal atresia repair, and regurgitation due to gastroesophageal reflux disease. Patients present with noisy breathing, barking or "TOF" (tracheo-oesphageal fistula) cough, recurrent respiratory infections, feeding difficulties, and apneic spells.[1] Mild to moderate cases generally resolve as the child grows and the cartilage strengthens. Severe cases, however, can have a mortality rate as high as 80%.[3]

Treatment of TM can involve symptomatic medical management, tracheal stents, which have little reported success,[4] and aortopexy. Aortopexy is currently considered the best option for the most severe cases and can be performed from a right, left, or anterior approach, using an open or thoracoscopic technique. Several criteria must be met before consideration for surgical repair. Once tracheomalacia is suspected as the cause of a child's respiratory distress, an awake direct laryngobronchoscopy (DLB) should be performed for definitive diagnosis. If TM is identified and it is intrathoracic, a computed tomographic angiogram can evaluate the relationship of the vasculature to the affected area. A dynamic computed tomography can help further isolate the specific region of the trachea that collapses as well as any surrounding structures that may aid in surgical planning.[5] Pre- and postoperative echocardiography evaluates the relationships and vascular integrity of the aorta and pulmonary arteries. If the workup reveals the aorta lying over the affected trachea, surgical intervention is warranted. Decisions regarding an open versus thoracoscopic approach, as well as where to enter the chest, will depend on the child's previous surgical history and imaging. Although we initially performed the thoracoscopic procedures via a left-sided approach,[6] we now prefer a right-sided approach to reduce possible complications from working near the pulmonary artery.

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Operative Technique

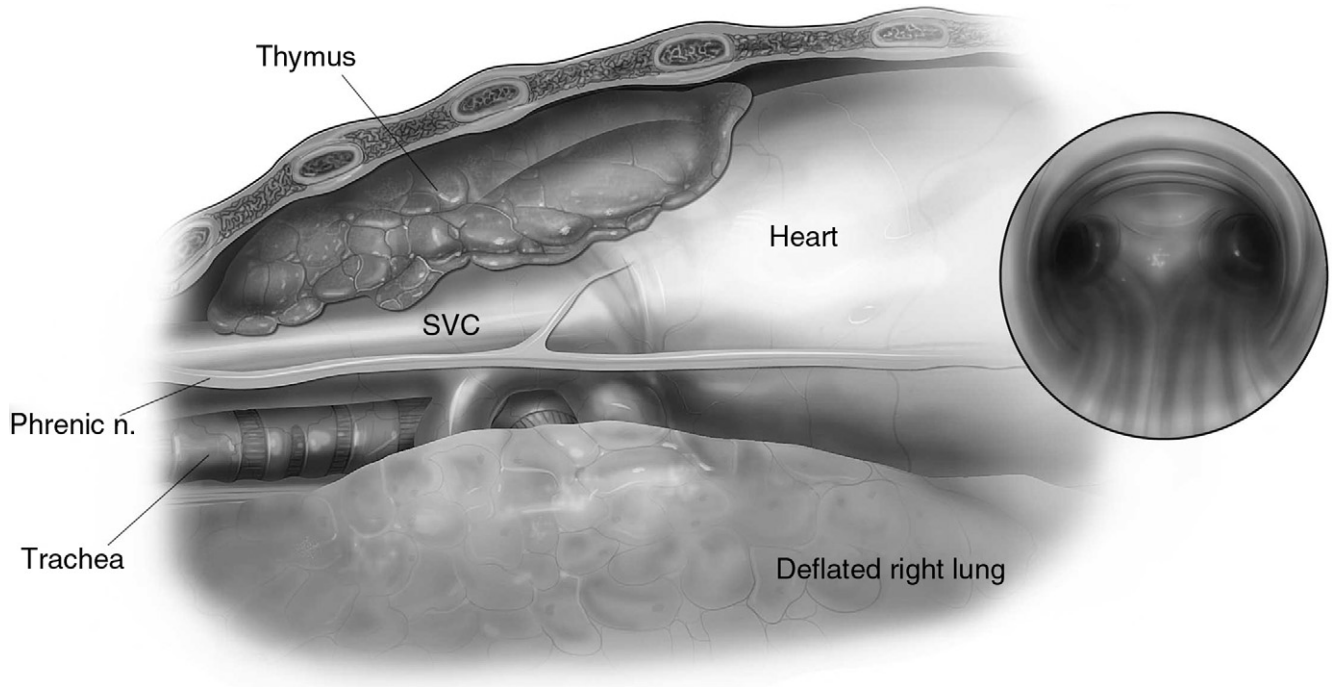

Figure 1 Normal tracheal anatomy with (inset) normal bronchoscopy. On arrival to the operating room, another DLB is performed while the child is breathing spontaneously to confirm once again that TM is present. This is an intrathoracic view of a normal airway with a firm and patent trachea. Bronchoscopy shows a trachea patent during both inspiration and expiration with no collapse. n. = nerve; SVC = superior vena cava.

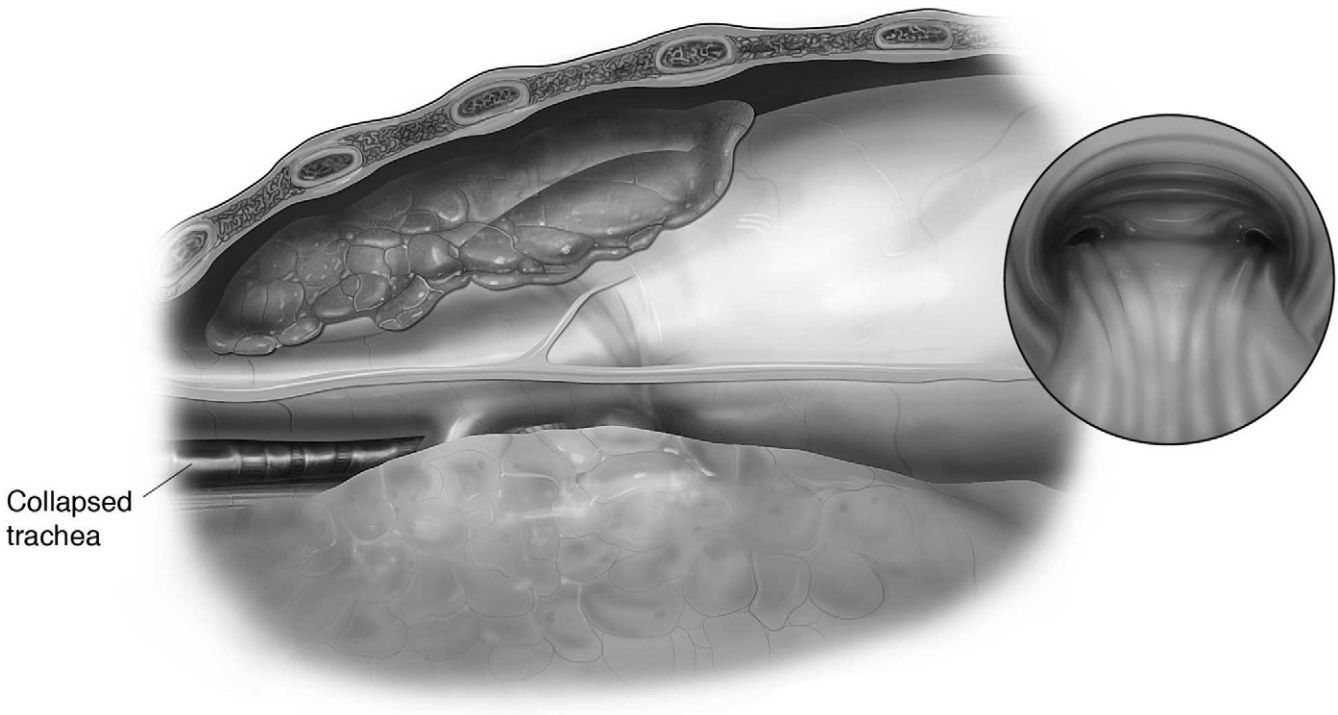

Figure 2 Tracheomalacia with (inset) tracheal collapse seen on bronchoscopy. Here, with TM, the weakened cartilaginous rings are collapsing under the great vessels. On bronchoscopy, the tracheal lumen is narrowed with expiration, revealing the obstruction underlying the symptomatology.

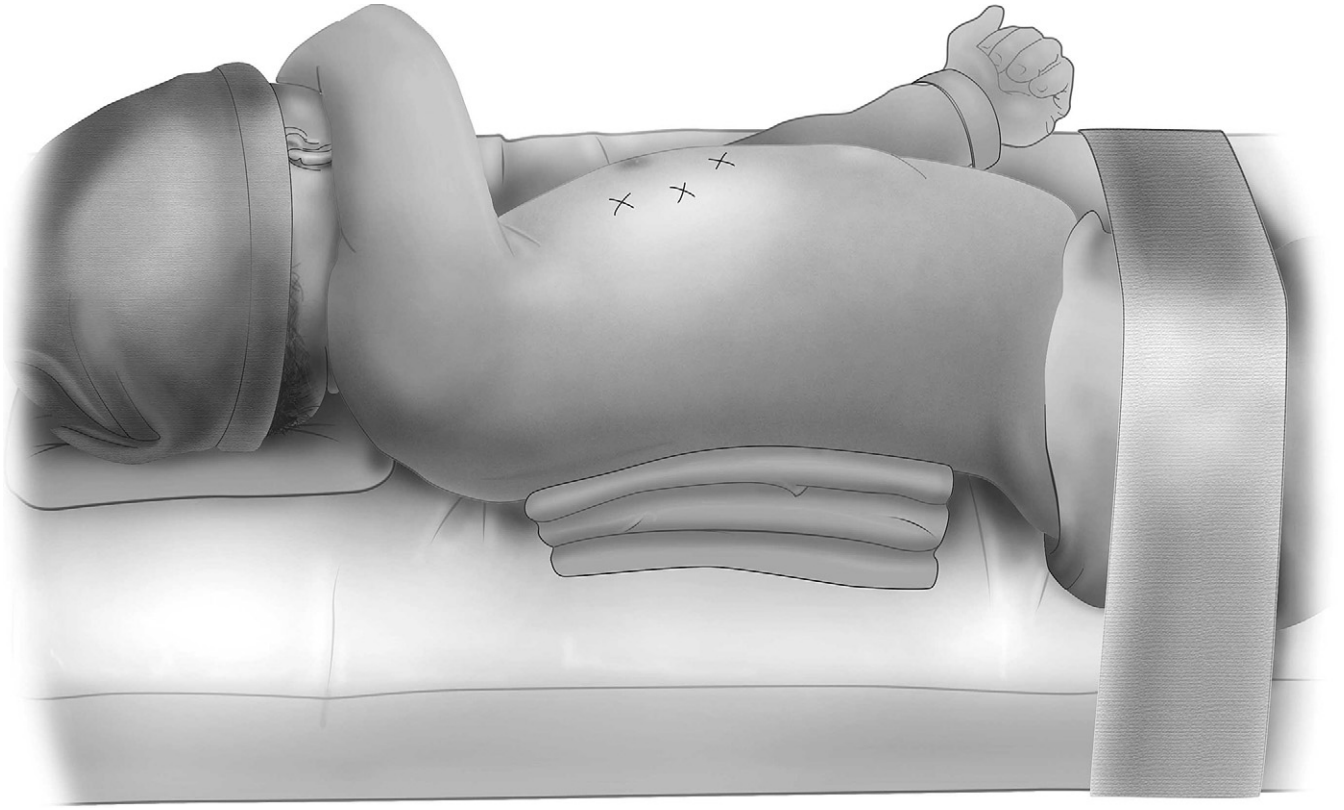

Figure 3 Patient positioning. The patient is positioned supine with the right side elevated to 20°-30°, to adequately expose the right anterolateral chest. The right arm is secured away from the operative field.

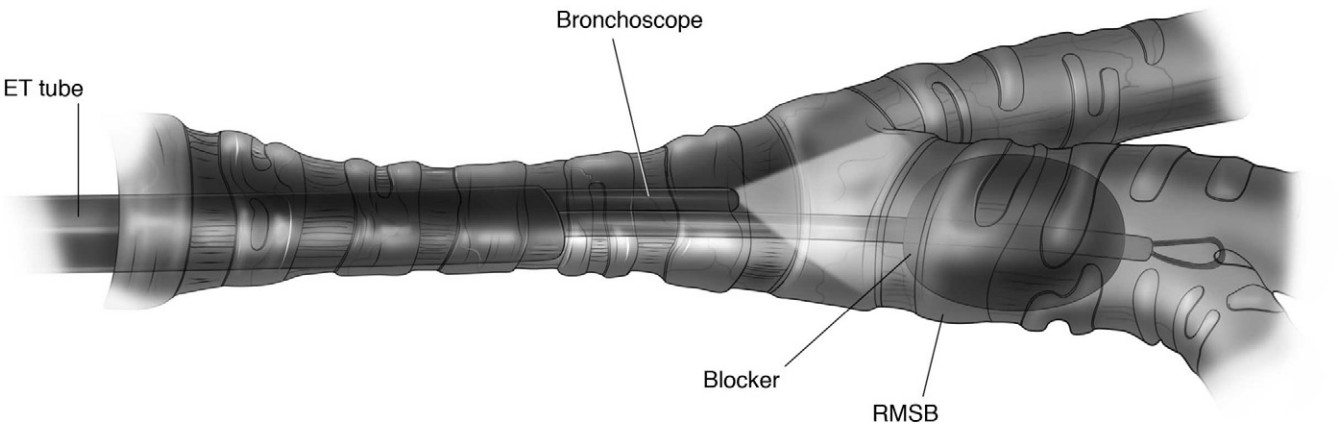

Figure 4 Bronchial blocker. Using a bronchial blocker inflated inside the right main stem bronchus, the right lung is deflated to create more exposure and working space within the right thoracic cavity. ET = endotracheal; RMSB = right main stem bronchus.

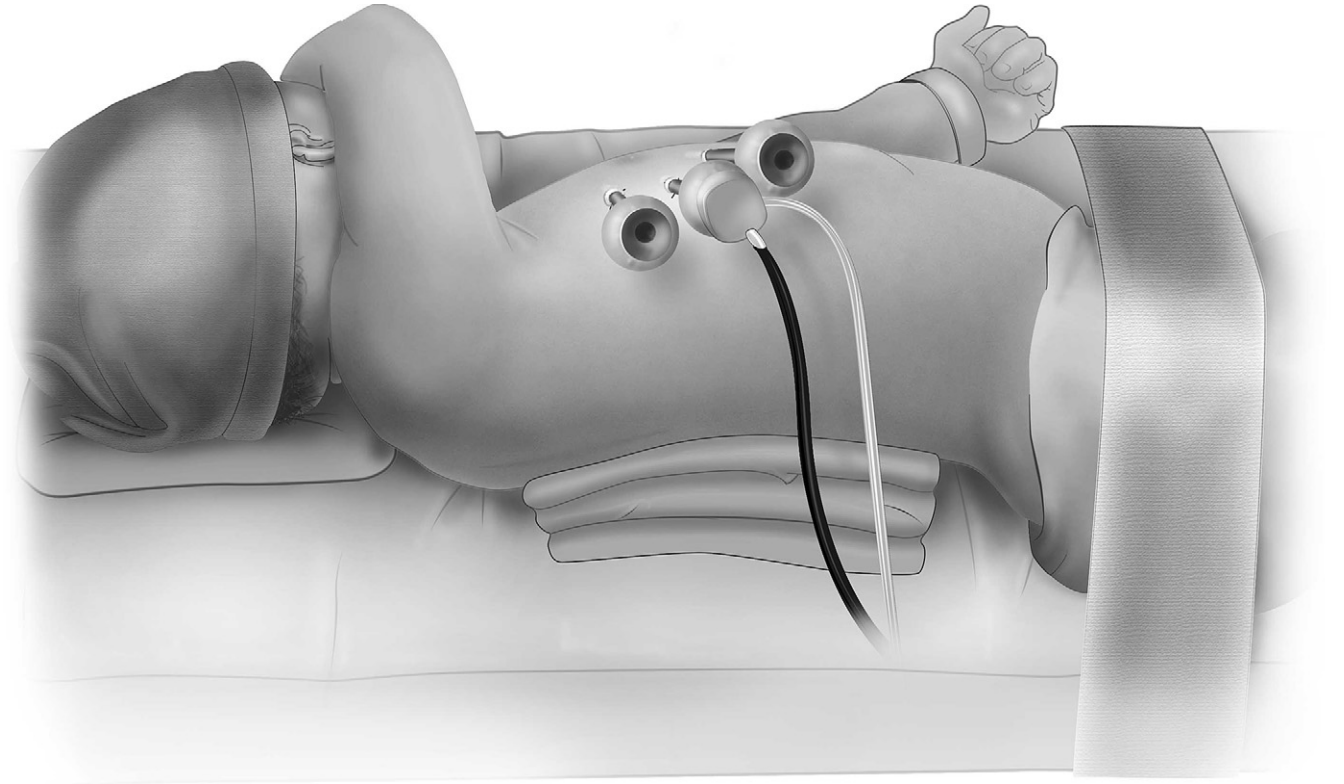

Figure 5 Trocar placement. Three- and 4-mm trocars are placed in the fourth and fifth mid-axillary and the sixth anterior axillary intercostal spaces. A 4-mm trocar is placed in the fifth intercostal space for the camera and 3-mm trocars are placed in the fourth and sixth intercostals spaces for instrumentation. Placement of these trocars should be such that they are aiming toward the manubrium. Adjustments to trocar location should be made as needed depending on the patient's anatomy. Insufflation pressures are set between 0 and 7 mm Hg.

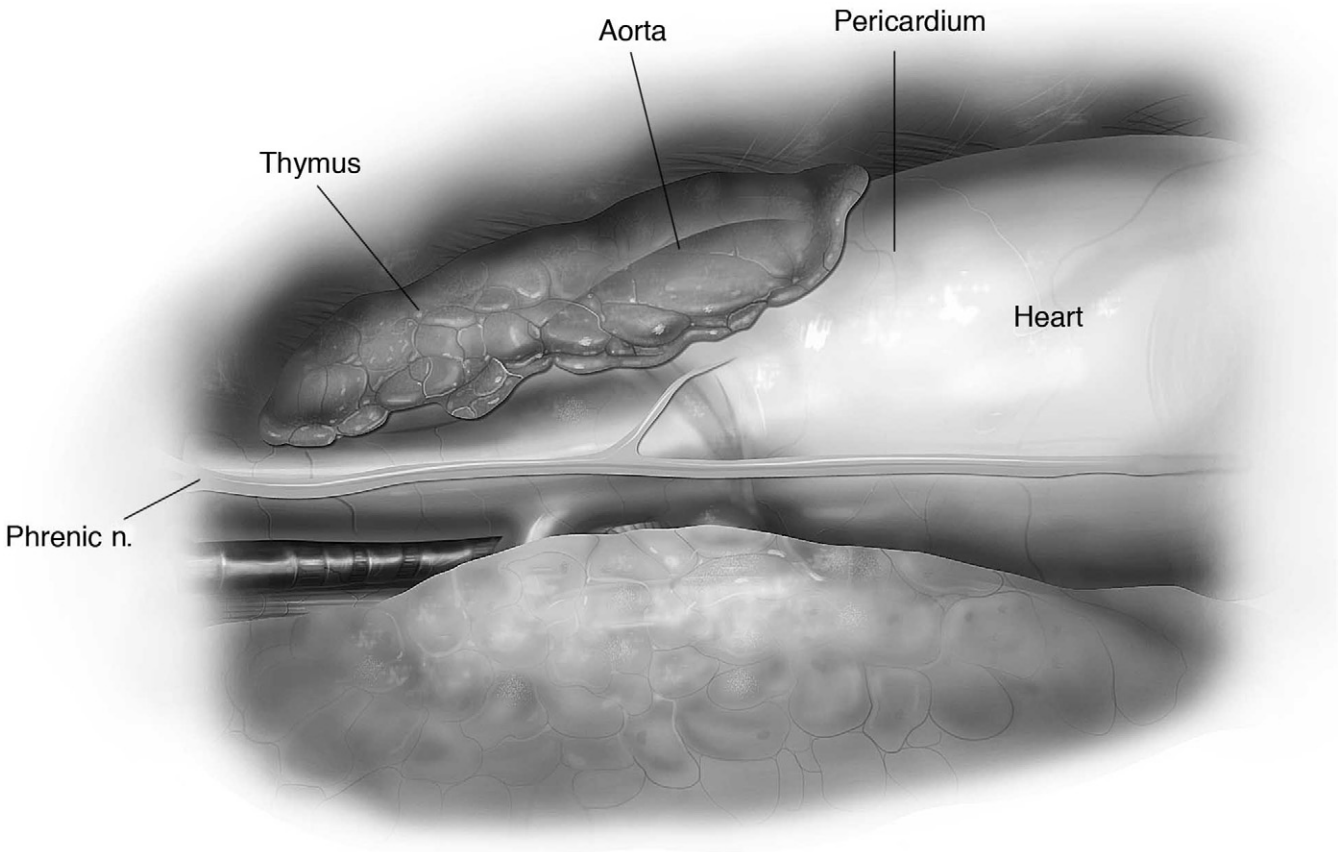

Figure 6 Initial view. On entering the right chest, the view consists mainly of the thymus, the heart, and the great vessels. The sternum can be seen partially obstructed by the large thymus. The phrenic nerve runs posteriorly along the pericardium; it should be identified and carefully preserved throughout the case. n. = nerve.

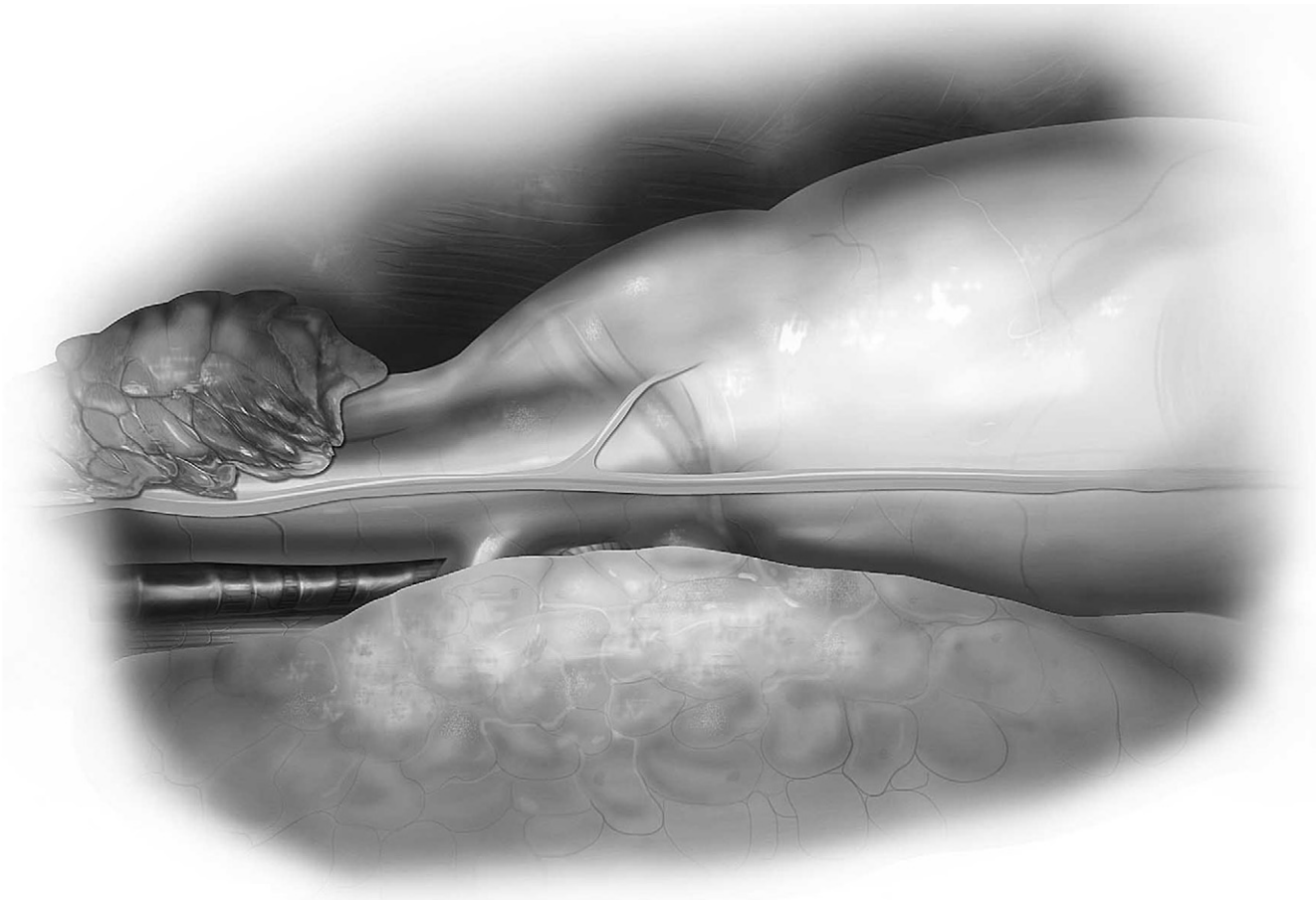

Figure 7 Thymus. The thymus is dissected free from the sternum and the pericardium and divided down the midline. The right lobe of the thymus is then swept superiorly into the apex of the right chest. The thymus can also be resected if adequate exposure is unattainable. This gives a more direct view of the aorta lying underneath the pericardium.

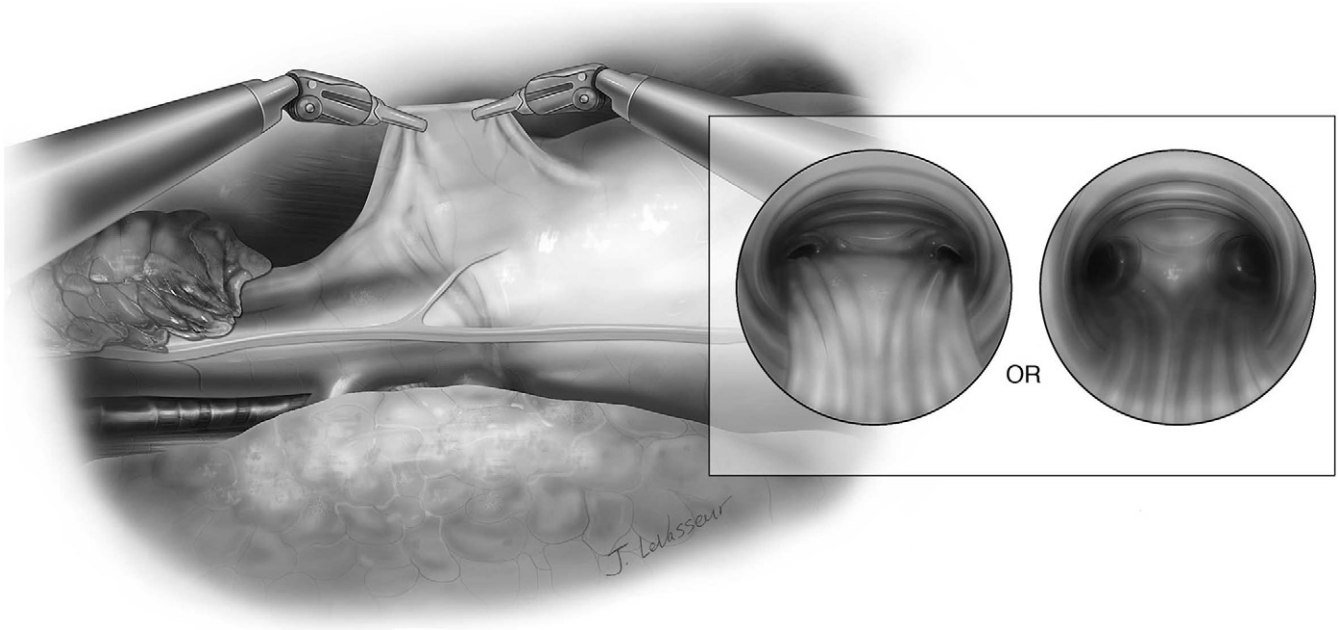

Figure 8 Pericardiopexy. In some cases, a pericardiopexy may adequately treat the TM and should always be attempted. The pericardium is lifted using 2 graspers and pulled toward the sternum. Simultaneously (inset), the bronchoscope is used to evaluate the degree of improvement in TM. This will be a more viable option in patients with less severe TM or TM that involves the distal trachea. If this allows acceptable tracheal opening, sutures are placed from the pericardium to the periostium of the sternum. The first suture is placed at the most superior aspect of the pericardium and work continues inferiorly. Another bronchoscopy is performed holding the sutures taut to confirm improved tracheal narrowing before securing the knots.

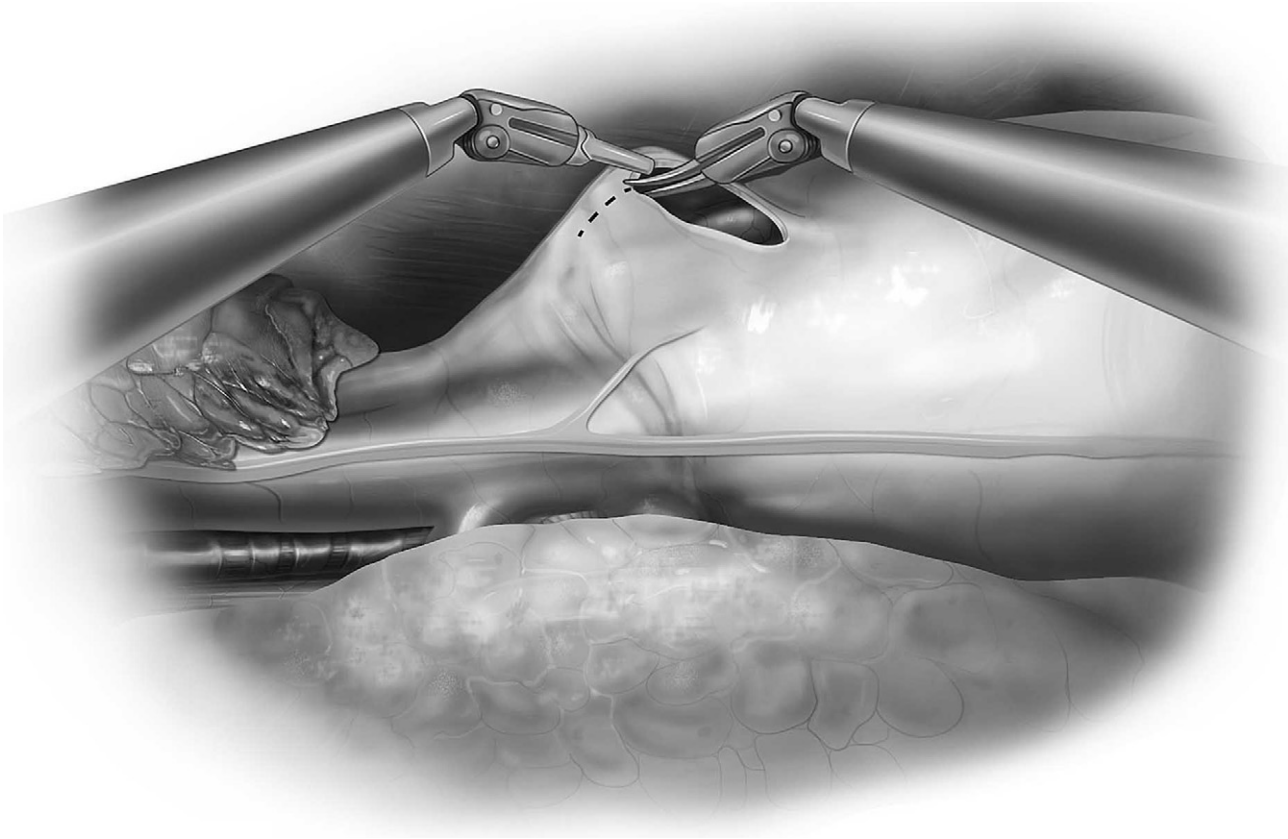

Figure 9 Opening the pericardium. If pericardiopexy does not provide adequate elevation or the affected trachea is too proximal, the pericardium needs to be opened to expose the underlying aorta. This can be done using the laparoscopic scissors, starting at the aortic root and extending as far as is necessary to attain adequate exposure.

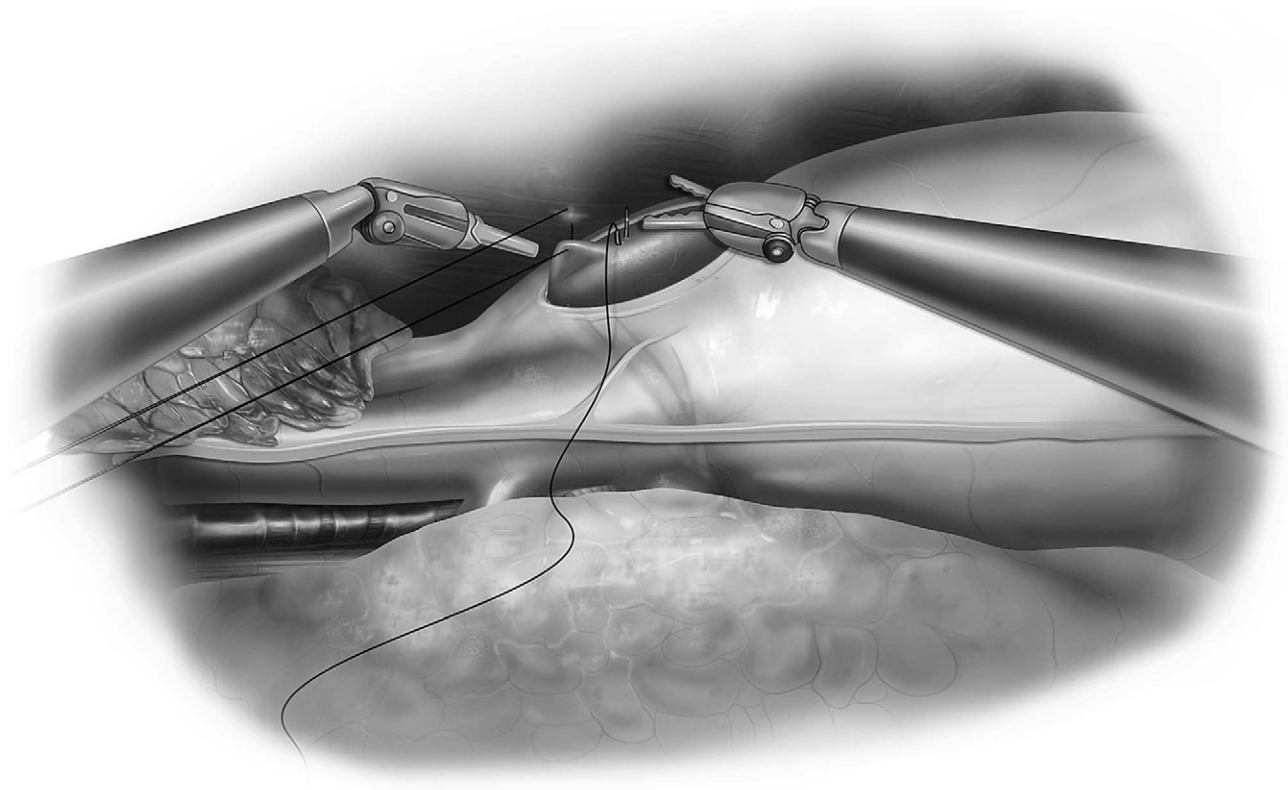

Figure 10 Aortopexy suture placement. Sutures are placed through the tunica media of the aorta and the sternal periostium. We use anywhere from 3 to 8 braided polyester sutures starting superiorly along the most anterior aspect of the aorta. It is important to place sutures precisely along the anterior aorta so that, once tied down, there is no aortic twisting. Sutures can be placed using a forehand or backhand technique depending on the surgeon's preference. If necessary, sutures may extend onto the innominate artery. It should also be noted that stitches placed too proximal (near the heart) may damage the aortic valve or involve the coronary arteries.

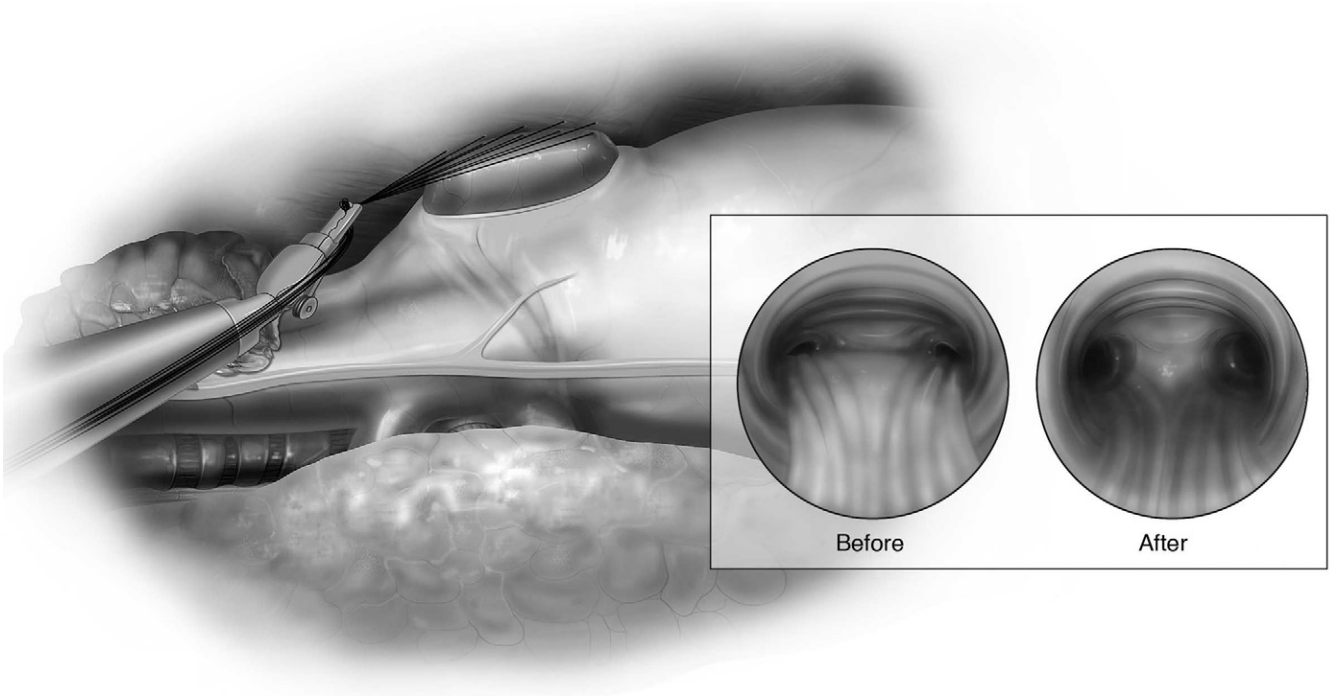

Figure 11 Bronchoscopic confirmation. Holding the sutures taut, bronchoscopy (inset) again confirms adequate tracheal opening before tying down the sutures. Once all sutures are secured, another bronchoscopy is performed.

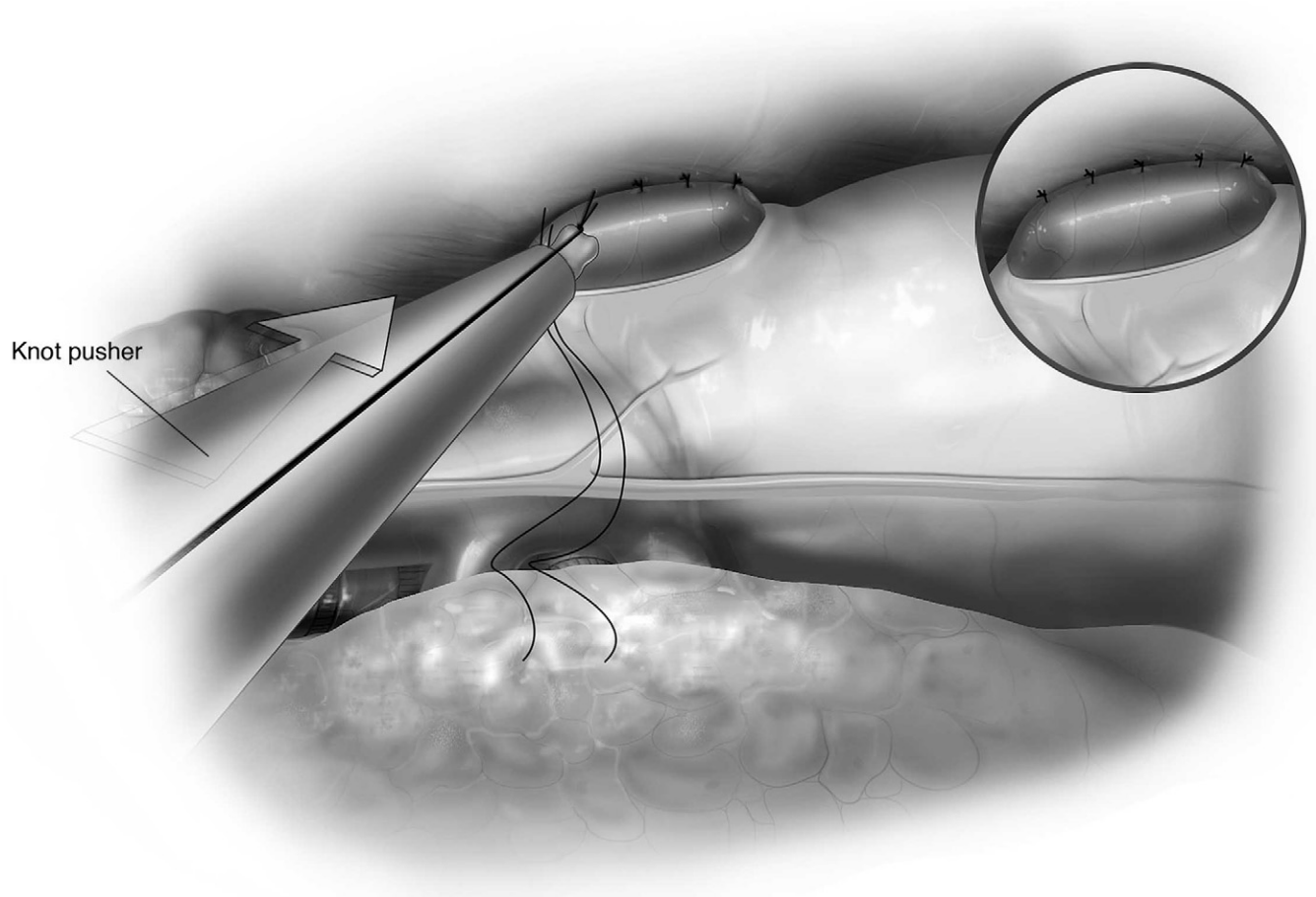

Figure 12 Knots and closure. Knots are secured down using a knot pusher. The inset shows the finished aortopexy with the aorta snugly against the sternum. A final DLB is performed with spontaneous breathing to confirm improved TM. If it was not removed, the thymus is replaced in a more natural position from the apex of the chest. A small chest tube can be placed through 1 of the port sites. The bronchial blocker is removed and the right lung is allowed to re-expand before closure. The remaining port sites are closed and sterile dressings are placed.

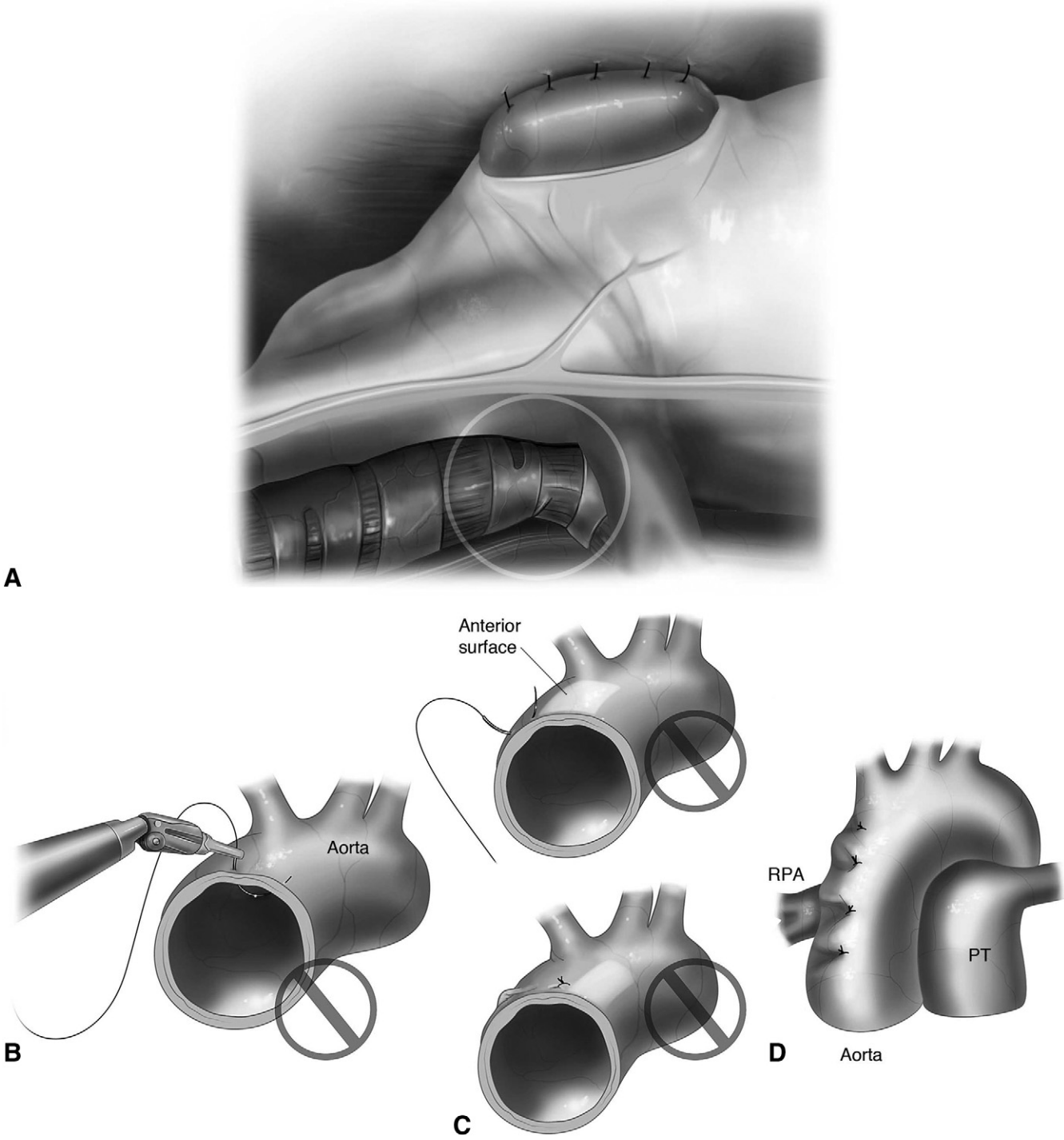

Figure 13 Things to avoid. (A) If there is too much anterior pull on the trachea, it can start to kink. On bronchoscopy, this will cause a narrowing in the anterior-posterior direction similar to that seen with TM. If this occurs, sutures will need to be removed and replaced with less anterior pull. (B) Sutures should be placed through the tunica media of the aorta. They should not enter the lumen as this causes profuse bleeding. (C) If sutures are not placed along the most anterior aspect of the aorta, there can be significant aortic twisting. Sutures are most likely to be placed too far along the right lateral aspect of the aorta, rotating it toward the patient's left. (D) Here is another view of the torqued aorta. The proximity of the right pulmonary artery is also visible here. It is of note that, although the pulmonary artery is less visible during a right-sided approach, it is still imperative to maintain awareness of its location and to avoid injuring it. PT = pulmonary trunk; RPA = right pulmonary artery.

Conclusions

Symptoms should be expected to improve quickly on recovery from anesthesia. Improvement after open aortopexy has been reported to be anywhere from immediately postprocedure up to 6 months postoperatively.[7,8] It should be noted that frequently some patients maintain some residual TOF cough and are persistently susceptible to upper respiratory infections. Generally, all patients need intensive care monitoring immediately following

surgery, but once extubated should be able to transfer to the ward shortly thereafter. The chest tube can be placed to water seal once the patient has recovered from anesthesia and can usually be removed on postoperative day 1 if there are no complications. We routinely perform a postoperative echocardiogram as previously mentioned. Discharge and follow-up are generally determined by the severity of any residual symptoms.

Compared with an open approach, thoracoscopic aortopexy has the potential for a quicker recovery with less postoperative pain. It can be performed safely and effectively with results similar to other surgical approaches. Possible complications from aortopexy with open thoracotomy include pneumothorax, phrenic nerve palsy, chylopericardium, atelectasis, and thymic engorgement causing lung compression.[7,8] Possible benefits of a thoracoscopic procedure include a better operative view, less postoperative pain and discomfort, improved cosmesis from smaller incisions, and better respiratory mechanics postoperatively.[6] Thoracoscopic aortopexy should, however, be reserved for straightforward cases of TM, with single segments of affected trachea identified as being amenable to improvement with manipulation of the great vessels. In patients with more anatomically complex TM or with tracheobronchomalacia, an open procedure, frequently through an anterior approach, is often more appropriate.

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