Thoracoscopic repair of esophageal atresia and tracheo-esophageal fistula in neonates: the current state of the art

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Abstract

The first thoracoscopic esophageal atresia with tracheo-esophageal fistula (EATEF) repair was performed in March of 2000. This report evaluates the results and evolution of the technique over the last decade. Thoracoscopic esophageal atresia repair has proven to be an effective and safe technique. Initial experience resulted in a higher stricture rate but this improved with experience and changes in technique over the last decade. The outcomes are similar to or superior to that of an open thoracotomy and avoid the musculoskeletal morbidity associated with that technique.

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Keywords

Tracheo-esophageal fistula
Esophageal atresia
Thoracoscopy

Introduction

Esophageal atresia (EA) with or without a tracheo-esophageal (TEF) fistula is one of the rarer congenital anomalies occurring in one in 3,000 births. Traditionally these patients have presented shortly after birth because of an inability to pass an oro-gastric tube, respiratory distress, or an inability to
tolerate feedings. The condition maybe associated with other major congenital anomalies (VATER syndrome), or may be an isolated defect [1–3]. Improvements in maternal-fetal ultrasound have resulted in prenatal diagnosis in a number of cases. This allows the surgeon to plan for delivery and eventual surgery. Patients with a tracheo-esophageal fistula require relatively emergent surgical intervention to prevent aspiration of gastric acid and over distension of the intestines. Those with pure atresia can be dealt with in a more leisurely fashion as long as the infants’ oral secretions are controlled by continuous or intermittent suction.

Advancements in technique and instrumentation in pediatric endoscopic surgery have allowed significantly more complex and delicate procedures to be performed, even in small neonates. Over the last 20 years, the number and breadth of minimally invasive surgical (MIS) procedures performed in infants have increased dramatically including the repair of esophageal atresia. [4–8]. In 1999, a stepping stone was laid when a successful thoracoscopic repair of a pure esophageal atresia was completed in a 2-month-old male [9]. In 2000, we reported on the first successful repair of an esophageal atresia with tracheo-esophageal fistula (EATEF) in a newborn using a completely thoracoscopic approach [10] and 2 years later reported on the first significant series [11]. These milestones allowed for a more widespread adoption of these techniques so that numerous pediatric surgical units around the world are now performing minimally invasive EATEF repair. There are now reports from over the world reporting excellent results using a thoracoscopic approach for this disease. This paper will review the development and technical advances of this technique as well as review the author’s personal experience.

**Technique**

The technique has been previously well described with mild variations between centers. A brief description will be given here for clarity. General endotracheal anesthesia is administered and an attempt is made to maintain low peak pressures until the fistula is ligated to prevent over distension of the abdomen. Single lung ventilation is not attempted, instead a low flow, low pressure of CO₂ (4 mmHg, 1 L/min) is used to collapse the right lung and create space.

**Positioning**
Once the endotracheal tube is secured, the patient is placed in a modified prone position with the right side elevated approximately 30º. If there is a right-sided arch then a left sided approach is used. This positioning gives the surgeon access to the area between the anterior and posterior axillary line for trocar placement, while allowing gravity to retract the lung away from the posterior mediastinum. This arrangement gives excellent exposure of the fistula and esophageal segments without the need of an extra trocar for a lung retractor. The surgeon and the assistant stand in front of the patient and the monitor is placed behind the patient. This allows the surgeon and the assistant to work in line with the camera towards the point of dissection. The assistant should not be placed on the opposite side of the table as this will place him at a complete paradox with the telescope. The scrub nurse can be on either side of the patient depending on the room layout. Because of the fine manipulation necessary the surgeon and the assistant should position themselves so that they are in the most ergonomic and comfortable position.

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Port placement

Port placement is extremely important because of the small chest cavity and the intricate nature of the dissection and reconstruction. The procedure can be performed with three ports but occasionally a fourth port is necessary to retract the lung.

The initial port (3–5 mm) is placed in the fifth intercostal space posterior to the tip of the scapula. This is the camera port and gives the surgeon excellent visualization of the posterior mediastinum in the area of the fistula and eventual anastomosis. A 30º lens is used to allow the surgeon to “look down” on his instruments and avoid “instrument dueling”.

The two instrument ports are placed to achieve a ninety degree angle at the presumed site of the anastomosis. The first port is placed in the mid-axillary line one to 2 interspaces above the camera port. This upper port is 5 mm to allow for the introduction of a clip applier and suture. A 3 mm port can be used if the surgeon wishes to suture ligate the fistula and passes the sutures through the chest wall. The lower port is 3 mm in size and is placed one or two intercostal spaces below, and slightly posterior to the camera port.
(Fig. 1). Ideally the instrument tips will approximate a right angle (90º) at the level of the fistula. This positioning will facilitate suturing the anastomosis. A fourth port can be placed either higher or lower in the thoracic cavity to help retract the lung, but this has not been necessary in the majority of cases. The operation then follows the same pattern as for the open procedure.

**Fig. 1**

Trocar placement with the infant in a modified prone position with the *left side* elevated 30º. The 30º scope is placed in the port just below and posterior to the tip of the scapula.

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**Ligating the fistula**

Once the chest has been insufflated and the lung collapsed, the surgeon must identify the fistula. In most patients, the fistula is attached to the membranous portion of the trachea just above the carina. This level is usually demarcated by the azygos vein.
After the azygos is identified, it should be mobilized for a short segment using a curved dissector or scissors. The vein is then cauterized and divided. Some advocate leaving the azygos intact, saying it improves vascularity of the area and decreases the leak rate. This point is still debatable but the vein may be preserved if desired.

With the vein divided, the lower esophageal segment is identified and followed proximally to the fistula. Because of the magnification afforded by the thoracoscopic approach it is easy to visualize exactly where the fistula enters the back wall of the trachea. A 5 mm endo clip can then be applied safely or the fistula can be suture ligated. Whichever technique is used, care should be taken to avoid the vagus nerve (Fig. 2).

**Fig. 2**
Exposure of the fistula for ligation

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**Mobilizing the upper pouch**

Attention is now turned to the thoracic inlet. The anesthesiologist places pressure on the NG tube to help identify the upper pouch. The pleura overlying the pouch are incised sharply and the pouch is mobilized with blunt and sharp dissection. The plane between the esophagus and trachea can be seen well and the two should be separated by sharp dissection. Mobilization of the upper pouch is carried on as far as necessary up into the thoracic inlet or neck depending on the length of the gap (Fig. 3).
Once adequate mobilization is achieved, the distal tip of the pouch is resected. This should be an adequate section so that there is a sufficient opening to prevent later stricture formation.

**The anastomosis**

With the two ends mobilized the anastomosis is performed using a 4-0 or 5-0 mono-filament absorbable suture in a single interrupted fashion. The back wall is placed first and then an Ng tube is passed under direct vision into the lower pouch and on into the stomach. The anterior wall is completed with the Ng tube acting as a guide to prevent incorporation of the posterior wall and ensuring patency of the anastomosis (Fig. 4).

**Fig. 4**

The completed anastomosis
Once the anastomosis is completed, a chest tube is placed through the lower trocar site and the tip is placed near the anastomosis (under direct vision with the endoscope). The other ports are removed and the sites are closed with absorbable suture (Fig. 5).

**Fig. 5**

Trocar skin incisions at the end of the procedure. A chest drain is placed in the left hand port
Author’s experience

From March 2000 to September 2012, 61 consecutive patients, 52 with esophageal atresia and a distal tracheo-esophageal fistula and 9 with pure esophageal atresia were referred to the author for repair. 16 had been diagnosed pre-natally and were delivered at the high risk, perinatal/neonatal center. Gestational age of the patients ranged from 30 to 40 weeks at the time of delivery. Three other infants with EATEF were also referred to the author during this period but were excluded because of size and associated anomalies. These three weighed 800, 1,100 g with a Tetralogy of Fallot, and 1,800 g with an omphalocele. In the thoracoscopic group, patients ranged in weight from 1.2 to 3.8 kg. Pre-operative evaluations revealed congenital heart disease in 22, including one patient with a Tetralogy of Fallot as well as a right-sided aortic arch. One patient had a double aortic arch that was right-side dominant. This patient underwent division of his left arch and repair of the EATEF during the same procedure. Three patients had a high imperforate anus and one had a cloaca. Six patients required intubation prior to surgery for increasing respiratory distress.

The gap length was estimated pre-operatively based on the position of the tip
of the NG tube and the apparent bifurcation of the trachea as seen on the CXR. This ranged from 2 to 4 1/2 vertebral bodies. At the time of surgery, the longest gap in a patient with EATEF was closer to 4 1/2 vertebral bodies as this patient had a trifurcation type fistula. The longest gap in a patient with pure atresia was 7 vertebral bodies.

Results

Sixty of 61 procedures were completed successfully thoracoscopically. The average operative time was 85 min (range 55–120 min). Esophageal contrast studies were obtained on postoperative day (POD)# 3 in 1 patient, POD#4 in 30, and POD# 5 in 28 patients and the anastomosis was patent with no evidence of a leak in 58/59. The first patient repaired with this technique had clinical evidence of a leak, saliva in the chest tube, on POD# 4. He was kept NPO and drainage stopped after 24 h. He was studied on day 8 with no evidence of a leak. Two patients with a long gap pure atresia also had clinical evidence of a small leak, which had sealed on a repeat study on day 9, and 10, respectively. The one conversion was performed in a patient with an unrecognized distal congenital esophageal stenosis. The anastomosis was successfully completed thoracoscopically but because the NG tube could not be passed into the stomach, the case was converted to ensure there was not a false passage.

Discussion

Many recent studies have documented the long-term outcome of patients with EA and TEF and the overall morbidity is significant, some related to the initial surgical approach and technique [12, 13]. The benefits of performing EA repair using minimally invasive techniques are obvious but the technical hurdles are significant. The greatest advantage is avoiding a postero-lateral thoracotomy in a neonate. This has been shown to be associated with a high degree of scoliosis and shoulder girdle weakness later in development [14–16]. A more recent study by Lawal et al. comparing children who underwent a thoracotomy vs. a thoracoscopic approach, showed a very high rate of scoliosis (54 %) vs. 10 % in the thoracoscopy group. They also did detailed measurements looking for chest wall asymmetry and found a much higher rate in the thoracotomy group. And lastly they polled the families
measuring for the satisfaction with the cosmetic result. They found a significant difference in the Manchester score between thoracotomy and thoracoscopy of 13.8 vs. 7.5. These data alone should be enough impetus for pediatric surgeons to push for a minimally invasive (MIS) solution for thoracic lesions in infants, and to avoid the musculoskeletal sequelae of a major thoracotomy (Fig. 6).

**Fig. 6**
Skin incisions one month post repair

The benefit of the improved cosmetic result of the thoracoscopic approach is actually something pediatric surgeons have valued and sought after for years. Bianchi and others have advocated muscle sparing and/or lateral thoracotomies, with various skin incisions to improve on this problem but these incisions can be difficult to develop, offer more limited access, and still require spreading of the rib interspace [17]. Despite an improvement over a standard thoracotomy incision, there are still significant issues associated with these various techniques. The cosmetic result is not comparable to that of a thoracoscopic approach, and may not eliminate the risk of scoliosis.

An unanticipated benefit of the thoracoscopic approach is the superior visualization of the anatomy and especially the fistula. Since the fistula is
visualized perpendicular to its insertion to the membranous trachea, the exact site for ligation can be identified easily, thereby minimizing the residual pouch attached to the trachea. The use of the 5 mm titanium clips has proven to be simple and effective with no evidence of tracheal leak or recurrent fistula. Others choose to suture ligate the fistula and this technique has been successful as well.

A recognized advantage after performing the first cases thoracoscopically was that the dissection and anastomosis were basically carried out in situ. Because the separation of the fistula and the upper pouch from the trachea was performed under direct magnified vision from a lateral approach there was little manipulation or force applied to the trachea itself. This may help diminish the degree of tracheomalacia that these children have post-operatively. Also the plane between the upper pouch and trachea was more obvious making injury to the membranous wall of the trachea less likely.

The improved visualization also helps with the mobilization. It is easy to see and dissect well up into the thoracic inlet allowing greater length to be obtained on the upper pouch in cases of a long gap. The distal end can also be visualized and mobilized more easily, especially in cases of pure EA. This has been borne out in our ability to anastomose primarily patients with gaps as long as seven vertebral bodies. By performing the anastomosis in situ there may be less tension on the esophageal ends allowing longer gaps to be brought together without tearing.

The major technical hurdle in this operation is the suturing of the anastomosis. The placement of the sutures and knot tying are technically demanding. Also as opposed to the open technique where the entire posterior row of sutures can be placed and then brought together to disperse the tension along multiple points during knot tying, this method places all the tension on one suture at a time. So far this has not been a significant problem but it could prove to be. For this procedure to become more widely accepted it may be necessary to develop a mechanical anastomotic device or self-knotting suture. The rate of anastomotic narrowing requiring at least one dilatation in our series was 20 % but was initially almost 50 %, a figure much higher than our open experience. This may have been secondary to inadequate approximation of the mucosal ends or an insufficient opening being made in the upper pouch. We have
modified the technique to eliminate both of these problems as well as switching to a monofilament absorbable suture. These changes seem to have resolved the problem in only 2 of the last 24 patients (10 %) requiring dilatation in the last half of the series [18].

There are now multiple studies from institutions all over the world comparing open vs. thoracoscopic EA-TEF repair. Lugo et al. [19] found the results and outcomes of a thoracoscopic approach comparable to that of the open. Tokhais et al. [20] also found comparable outcomes in a multi-institutional study. A recent survey, performed by IPEG (International Pediatric Endosurgical Group), of its members showed wide adoption of the thoracoscopic approach. 170 surgeons responded from 31 countries and over half stated that a thoracoscopic approach was their first choice [21]. This shows that the technique is not just limited to “the experts” and that many centers which practice advanced MIS have been able to adopt and master the technique.

Perhaps the most compelling paper is a recent review by Borruto et al. [22] in which they reviewed 5 large series that compared open vs. thoracoscopic repair and performed a meta-analysis. They showed no statistically significant difference in the complications or outcomes between thoracoscopic vs. the open repair. (Table 1) This would mean the major difference is the avoidance of a major thoracotomy, the advantages of which have already been discussed.

Table 1

Comparative studies of open vs thoracoscopic TEF showing no significant differ outcomes or complications

<table>
<thead>
<tr>
<th>Author</th>
<th>Type of study</th>
<th>Level of evidence</th>
<th>No. of thoracoscopies vs. no. of thoracotomies</th>
<th>Endpoints</th>
<th>Results thoracoscope vs. thoracotomy (%)</th>
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<tbody>
<tr>
<td>Borruto et al.</td>
<td>Meta-analysis</td>
<td>3a</td>
<td>69 vs. 97</td>
<td>leakage rate</td>
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There has been concern about the physiologic stresses during a thoracoscopic repair because of the CO2 insufflation, and one lung ventilation. Some are concerned that there may be significant hypercapnia, acidosis, and cerebral hypoperfusion. A recent study from Bishay et al. [23] from London looked patients undergoing thoracoscopic CDH and TEF repair vs. those undergoing a thoracotomy. In the CDH group they found significant evidence of hypercapnia and acidosis in the thoracoscopic group. They found similar findings in the TEF group but these did not reach statistical significance. However, other studies including two recent reports at the EUPSA and IPEG meetings by Stolwijk from Utrecht showed no such problem [24, 25]. They measured for hypercapnia, acidosis, and cerebral perfusion and found no significant difference between the open and thoracoscopic group. They did use average pressure of 5 mmHg which is significantly less than that of the London group and their operative times were less and perhaps this explains the results of the other study. In any event many other series including our own
have not shown clinical problems of significant hypercapnia or acidosis in these patients.

Clearly the technical and physiologic hurdles to accomplish this type of repair are many and it will require continued advances before this surgery becomes commonplace. However, the ability to perform this complex reconstruction without a thoracotomy lays further ground work in minimizing surgical morbidity in even the smallest pediatric patients.

References


less than 5 kg: experience of 649 cases. Surg Endosc 22:2214–2219


