Guidelines of the Italian Society of Videosurgery in Infancy for the minimally invasive treatment of the esophageal atresia

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Background

The SIVI (Italian Society of Videosurgery in Infancy) guidelines are clinical practice guidelines edited and approved by the Society’s steering committee. They are the products of a detailed systematic review of the literature, integrated with expert opinion in the field of pediatric minimally invasive surgery.

These guidelines are intended to assist the pediatric surgeons experienced or not experienced in minimally invasive surgery, with the goal to inform choices related to the indications, approach, and techniques to use when treating the major pediatric surgical pathologies.

Given the complexities of congenital malformations and other pediatric surgical conditions, as well as large variations in available regional health services, we must note that these guidelines are not intended as a cookbook recipe to follow for all possible patients. Rather, the guidelines should serve as a flexible framework, to be used by the physician in concert with the parents, to choose the best approach for each individual patient. Decisions tailored to available scientific knowledge and the needs and desires of the patient’s family serve both patient autonomy and medical science.

All guidelines are published in this scientific Journal, in order to ensure their availability to all physicians.

The Guidelines project has been approved by the SIVI General Assembly of the 2016 Madrid congress.

Review of guidelines has been performed by the Steering Committee of SIVI and experts.

Introduction

Esophageal Atresia (EA) is defined as an interruption in esophageal continuity that results in a proximal tract that ends in a blind pouch in 98% of cases, and a distal tract that in 87% of cases arises via a Fistula from the Trachea (TEF). This malformation is caused by an abnormality in the process of separation of the trachea and the esophagus that normally occurs during the fourth week of gestation. Currently, a specific genetic anomaly is detected in only 11-12% of patients; most cases of EA/TEF are sporadic. The recurrence risk is very low (~1%), which suggests that epigenetic and environmental factors contribute to the development of EA/TEF.

There are several classification schemes proposed to distinguish the various types of defect: the Gross scheme is purely anatomic, and correlates with incidence (Table 1); the Spitz classification combines EA/TEF with the patient’s Birth Weight (BW) and the existence of concomitant cardiac anomalies (Table 2); it has prognostic value, and is the most commonly used scheme today.

Diagnosis

Prenatal Diagnosis

Suspicious findings on prenatal ultrasound are: polyhydramnios (if isolated finding, PPV = 35-63%; if associated with small or absent gastric bubble, the PPV increases to 67%); small or absent gastric bubble; abnormal fetal swallowing movements; dilated proximal esophageal pouch (visible after the 23rd week of gestation); or, polyhydramnios associated with a constellation of

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fetal anomalies such as VACTERL (Vertebral defects, Anal atresia, Cardiac malformations, TEF, Renal or Limb anomalies).

When the ultrasound is uncertain, fetal Magnetic Resonance Imaging (MRI) may be diagnostic, and it also allows a detailed investigation of associated abnormalities.

The prenatal diagnosis does not affect the prognosis of any given patient, but early diagnosis provides the advantage of appropriate prenatal counseling, as well as the expeditious planning of delivery and postnatal care in a well-equipped tertiary care center.

There are no inherent indications to anticipate delivery of a fetus affected by EA/TEF, nor does the baby benefit from cesarean delivery.

Postnatal Diagnosis

In patients where EA/TEF is suspected prenatally, the placement of a nasogastric tube will rapidly confirm the diagnosis.

Symptoms in the newborn may raise suspicions in the first hours of life, even when there has been no prenatal diagnosis: salivory (unusual in premature patients); cough and regurgitation with the first attempts at feeding; cyanosis, associated or not with feedings, due to aspiration of saliva or milk; respiratory distress; aspiration pneumonitis may rarely occur due to passage of gastric secretions into the trachea via the TEF.

The following findings should raise immediate concern for EA/TEF:
- First, placement of a Nasogastric (NG) tube into the proximal esophagus, with inability to pass into the stomach (mean distance from the mouth to the cardia in a term neonate is 17 cm).
- Second, a radiograph that shows the NG tube curled in the proximal esophagus, confirmed with a small amount of contrast material. The radiograph may also confirm distal TEF by demonstrating air in the stomach and intestine; the use of contrast permits better visualization of the length of the proximal esophageal stump, as well as the identification of the rare proximal TEF.
- Third, evaluation of the esophageal gap. In patients with EA without distal TEF (Gross types A and B), it is useful to measure the gap between proximal and distal esophageal stumps. This measurement may be delayed by 15 days; in the meantime, a gastrostomy may be placed to ensure adequate enteral nutrition. To measure the esophageal gap, and, more importantly, the mobility of the distal stump, the surgeon may introduce a 4-French Hegar dilator via the gastrostomy, pushing it cephalad. In this way, the gap may be measured accurately; the dilator may also be connected to a dynamometer that can measure the elastic force of the distal stump. The force needed to stretch the stump is proportional to 200-300 grams. The gap should be measured fluoroscopically, with and without pressure. Patients treated with prior esophagostomy may have similar measurements performed from above, assisted by a radiopaque marker at the stoma. Preoperative gap distances may be helpful to the surgeon in choosing the timing of operative repair.

Preoperative Management

Since many EA/TEF patients have associated anomalies, the preoperative evaluation should include a careful physical exam, with particular attention to the external genitals and the anus; echocardiography; renal and urinary tract ultrasound; and brain ultrasound.

Patients should be cared for in the Neonatal Intensive Care Unit (NICU) in a semi-sitting position to minimize the risk of aspiration of saliva or gastric secretions via the TEF; NPO (‘nothing by mouth’) with total parenteral nutrition; avoidance when possible of mechanical ventilation or Continuous Positive Airway Pressure (CPAP) to prevent gastric distension (distal TEF in most patients), which may worsen the preoperative respiratory status.

Surgical Approach

The goal of surgical correction is to close the TEF and to reconstruct a complete patent esophagus via an end-to-end anastomosis of the proximal and distal esophageal stumps. This goal may be reached using different operative approaches.

Preoperative Bronchoscopy

There is still debate about the need for preoperative Tracheobronchoscopy (TBS) in patients with EA/TEF: a recent European survey showed that only 43% of pediatric surgeons regularly utilize TBS prior to surgical correction. When used, it has several roles: to evaluate vocal cord function; to search for associated airways anomalies such as tracheobronchomalacia or clefts; to define the esophageal gap; and, to confirm the presence and location of TEF. When identified, the TEF may be cannulated with a small catheter (i.e., Fogarty) which serves as a landmark during surgical repair. To accurately measure the esophageal gap in patients with a distal TEF, a radiopaque catheter is placed in the proximal pouch at the same time that the tip of the bronchoscope is positioned at the TEF. Fluoroscopy may then be used to determine the gap.

Thoracotomy Approach

Informed Consent

The surgeon should discuss with the parents: the various surgical approaches and their risks and benefits; immediate postoperative management in the NICU, and the possible presence of thoracostomy tube, NG tube, and central venous catheters; intraoperative risks of bleeding or inability to close the esophageal gap in a single procedure; postoperative risks such as pneumothorax, anastomotic dehiscence leading to empyema or mediastinitis; stenosis of the anastomosis, or persistent TEF or tracheal leak; and, late complications such as tracheomalacia or esophageal dysmotility.

Operating Room Setup

After induction of general anesthesia via endotracheal intubation and the placement of appropriate monitors, IV, and arterial
access, the neonate is positioned in the left lateral decubitus position, with the right arm raised above the head. The first surgeon stands at the patient’s back, on the right side of the table, with the assistant on the left.

**Surgical Technique**

The traditional surgical approach is via a right posterolateral thoracotomy, unless preop echocardiography has demonstrated a right-sided aortic arch, when the incision should be on the left. The skin incision starts about one centimeter below the inferior pole of the right scapula, follows along the rib, and ends at the midaxillary line. After dividing the latissimus dorsi, the surgeon then incises the 4th intercostal muscle along the superior border of the 5th rib to enter the right hemithorax. The intact parietal pleura is peeled off the thoracic wall to gain extra-pleural access to the posterior mediastinum. At this point, the surgeon may choose to ligate and divide the azygous vein (usually found at the level of the distal TEF); then, the distal esophageus and TEF are identified, and, with the anesthesiologist gently pushing down on a nasogastric tube, the proximal pouch is also located. The distal TEF is ligated and then divided from the trachea; afterward, gentle blunt dissection is performed to mobilize the proximal and distal esophageal stumps. Traction sutures (5-0 or 6-0) may be placed in both stumps to aid in the mobilization, and to minimize handling of the terminal anastomotic tissues. Once the stumps have been freed, the proximal esophagus is opened, and an end-to-end anastomosis is fashioned from the proximal opening and the distal esophageal stump, using interrupted sutures placed around the NG tube, which has now been extended into the stomach. When the anastomosis closure is complete, a perianastomotic drain is positioned. At the end of the procedure, the thoracotomy is closed without costal synostosis, with the perianastomotic drain on Boula. The skin is closed with absorbable subcutaneous 5-0 or 6-0 sutures.

Closure of the TEF and the end-to-end esophageal anastomosis are usually performed in a single procedure. In cases of long gap EA, after an accurate dissection and mobilization of both stumps, the proximal stump may be mobilized even further via a cervical incision if there is too much tension to successfully complete the esophageal anastomosis. After this maneuver, if the distance to bridge the gap is still too large, the surgeon may apply gentle traction to both stumps for 10-15 minutes. If the stumps still don’t reach, other techniques that may permit a successful anastomosis are the creation of a proximal esophageal flap, or the Foker/Kimura technique of internal and external traction.

**Postoperative Management**

The patient is transferred directly from the operating room to the NICU when the intervention is complete, and remains intubated, sedated, and paralyzed for the first 24-48 hours postop. The surgeon may desire to maintain pharmacologic paralysis for a longer period of time if more tension was needed to close the esophageal gap. After approximately 48 hours, if the patient is doing well clinically, then paralytic agents are stopped, and the patient may be extubated. Between postop days 6-8, radiologic evaluation of the esophagus is performed with water-soluble contrast material to rule out anastomotic dehiscence or stenosis. Feedings by mouth may be slowly initiated at this time if there are no contraindications. By postop days 8-9, the trans-anastomotic NG tube and the perianastomotic drain may be removed if there is no evidence of stenosis or of leakage at the site of the anastomosis. Nutrition in the postop period is obtained via TPN through a central venous catheter and may be progressively diminished as the patient’s feedings by mouth are advanced. Broad spectrum antibiotic coverage is provided, as well as maintenance analgesia in the immediate postop period, which can be decreased to a PRN basis after several days.

**Minimal Access Thoracotomic Approach**

In order to reduce complications related to an open thoracotomy procedure, the traditional surgical approach has been modified over the years. The mini-invasive thoracotomy approach (minimal access surgery, or MAS) differs from traditional thoracotomy in the initial surgical incision: the skin incision follows an axillary skin fold, and extends from the posterior axillary line to the anterior axillary line, as high up as possible while still permitting entry into the thorax (3rd-4th intercostal space). Instead of dividing the underlying muscles, the fibers are gently separated and retracted above and below the incision (muscle sparing technique). The dorsal thoracic neurovascular bundle and the long thoracic nerve are identified and conserved. The serratus anterior muscle is separated parallel to its fibers to expose the ribs. Access to the hemithorax is obtained subperiosteally, thus also sparing the intercostal muscle. The intrathoracic part of the procedure continues as described above for the traditional thoracotomy.

The smaller subaxillary incision offers better aesthetic results, since the scar migrates upward into the axilla with somatic growth. The nerve conservation and muscle sparing techniques reduce the risks of right hemithoracic deformities, scoliosis, and winging of the scapula.

**Thorascoscopic Approach**

**Informed Consent**

In addition to the usual risks, benefits, adverse effects, and complications as described above, the surgeon must also: review the different options of surgical approach available; discuss the technical aspects of Video-Assisted Thoracoscopy (VATS); and discuss the eventual reasons to convert a mini-invasive thorascopic procedure into an open thoracotomy, if necessary.

**Operating Room Setup**

After induction of general anesthesia via endotracheal intubation and the placement of appropriate monitors, IV, and arterial access, the neonate is positioned in the left lateral decubitus position with 30 degrees prone, with the right arm raised above the head. To maintain the correct position, the right iliac crest may be immobilized with adhesive tape. The patient should be stabilized along the left edge of the operating table. The lead surgeon stands on the left side of the table, in front of the patient, with the assistant adjacent to the left. The scrub nurse and monitors are placed behind the patient, on the right side of the table.

**Surgical Technique**

Thoracoscopy is performed through a trans-pleural approach, and usually three access ports are inserted for instruments: a 5 mm trocar for 30 degree optic camera is placed about one cm below and laterally to the inferior pole of the right scapula; a 3-5 mm operative trocar is placed in the midaxillary line between the 2nd and 4th intercostal space; and, a second 3 mm operative trocar (rarely 5 mm) is placed along the posterior axillary line, between the 7th and 8th ribs.

The procedure commences with CO2 insufflation of the right pleural space to a pressure of 6-8 mm Hg; once the right lung is collapsed, the pressure may be reduced to the minimal needed to maintain lung atelectasis and permit visualization of the surgical field. The next step is to identify the azygous vein, which is usually ligated and divided to provide easier access to the esophageal.
stumps in the posterior mediastinum. (An azygous-sparing technique has also been described, where the esophageal anastomosis is created posterior to the intact vein.)

The TEF is then located, and is closed by either transfixing sutures, or by titanium endoclips, so the site may be seen by plain x-ray in the post op period. The TEF is then divided from the trachea, and the presence of a leak is evaluated using a “water test”. The two esophageal stumps are then mobilized, the proximal stump is opened inferiorly, and the end-to-end esophageal anastomosis is completed using between 8 and 12 interrupted 5-0 endoscopic reabsorbable sutures. After the suturing of the posterior wall, and before completing the closure of the anastomosis, the anesthesiologist places an adequate transanastomotic nasogastric tube into the stomach. At the end of the procedure, the operative trocars are removed under direct vision (to rule out thoracic wall hemorrhage), and usually, a perianastomotic drain is positioned. The trocar incisions are then closed in layers with reabsorbable sutures, using 5-0 subcutaneous sutures for the skin.

Postoperative Management

Postoperative management is identical to that described for the thoracotomy approach, although thoracoscopic patients usually require less postoperative analgesia, due to the much smaller surgical incisions.

Contraindications to Thoracoscopy

The main contraindication is lack of operator experience in the thoracoscopic repair of EA/TEF. Rarely, the patient has ventilation problems during thoracoscopy; if the anesthesiologist cannot adequately ventilate the patient during pleural insufflation, the procedure may need to be converted to an open approach.

Conclusions

The pediatric surgical literature has demonstrated over the past several years that thoracoscopic repair of EA/TEF is rapidly becoming the standard of care for these patients, not only for aesthetic reasons, but also because the short and long-term outcomes appear to be superior.

One area of contention relating to thoracoscopic procedures in the neonate is the physiologic basis on which the procedure depends: insufflation of CO₂ into the pleural cavity may cause hypercapnia and acidosis even in the absence of hypoxia or poor tissue perfusion, and these effects are proportionally greater in the neonate. The long-term effects of these acid-base shifts on brain development are unknown, and this has fueled debate between the two proponents, about the relative merits and risks of open thoracotomy versus thoracoscopy. Existing studies have not shown significant elevations in hypercapnia or acidosis that would be concerning during any other surgical procedure without CO₂ insufflation; neither have any follow-up series of patients already operated upon via the thoracoscopic approach demonstrated an increase in late neurologic complications. It should be noted that due to the bony thoracic cage, only a very small amount of CO₂ need be insufflated to collapse the lung for the procedure, thus minimizing the risk of significant hypercapnia.

Compared to open thoracotomy, the thoracoscopic approach offers several technical advantages: slow and gentle CO₂ insufflation allows the gentle collapse of the right lung, thus avoiding trauma to the pleural and the parenchyma that necessarily occurs during pleural peeling; magnification from the optics permits better visualization for the surgeon, especially helpful in the esophageal stump mobilization steps of the procedure, in identifying nerves and small arteries that are better conserved, and in localizing the TEF and closing it without a leak.

Recent studies have shown how the thoracoscopic approach provides other advantages as well, such as decreased postop intubation time, shorter time to start and full advancement of oral feeding, and decreased duration of postop analgesia. Therefore, inpatient care for these patients is tending to decrease in duration, ultimately reducing the overall costs for EA/TEF repair in the neonate.

Thoracoscopic repair of EA/TEF does not appear to increase the incidence of significant gastroesophageal reflux in the long term, and the excellent aesthetic results with minimal scarring may be added to the decreased risk of thoracic wall deformities or scoliosis.

Additionally, there are increasing numbers of patients who are operated upon using the thoracoscopic approach despite being preterm or low birth weight, and the results do not appear inferior to the open procedure. If necessary, a second thoracoscopic procedure may be performed without added complications.

The main limit to the wider use of thoracoscopic repair of EA/TEF is the technical complexity of the procedure, that requires a high level of surgical experience; this often translates into longer operating times compared to open thoracotomy, and into the need for a minimally invasive surgical multispecialty team to care for these patients (neonatologist, surgeon, anesthesiologist).

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