39 Anesthesia for Tracheoesophageal Fistula

Pornswan Ngamprasertwong

Introduction

Tracheoesophageal fistula (TEF) with or without esophageal atresia (EA) is a rare congenital anomaly, with an incidence of 1:2500–3000 live births. The majority of cases are sporadic, but can be associated with VACTERL (vertebral defects, anal atresia, cardiac defects, tracheoesophageal fistula, renal anomalies, and limb abnormalities) [1]. There are two common classifications, by Gross and Vogt, using anatomical structure to classify this anomaly into five different types. The most common type is Gross type C (Vogt type IIIb), which is EA (blind upper pouch) with a distal fistula (TEF of the lower pouch). Typically, TEF with EA patients require surgical intervention in the very first few days of life. The TEF without EA patients (Gross type H) might be undetected during the neonatal period, but generally present later on with recurrent aspiration and pneumonia. For isolated EA (Gross type A), patients are managed by gastrostomy, follow by re-anastomosis in the next few months. Due to an intact tracheal lumen, the isolated EA poses less respiratory and anesthetic challenges. The summary of anesthetic plan with reasoning is demonstrated in (Table 39.1).

P. Ngamprasertwong, M.D., M.Sc
Department of Anesthesia and Pediatrics, Cincinnati Children's Hospital Medical Center,
Cincinnati, OH, USA
e-mail: pornswan.ngamprasertwong@cchmc.org

Pathophysiology and Clinical Features

In contrast to many other congenital anomalies, TEF is frequently missed prenatally despite the many advances in prenatal diagnosis. Polyhydramnios and the absence of a stomach bubble in the prenatal ultrasound are nonspecific signs of EA with a positive predictive value of 56% [1]. In EA with TEF patients, air in the stomach may be present and the finding on the prenatal ultrasound is less specific. The diagnosis of EA with TEF is usually made shortly after birth based on clinical symptoms including: coughing, wheezing, excessive salivation, choking, cyanosis with feeding, and the inability to pass a nasogastric tube more than 10–15 cm. Definitive diagnosis is confirmed by coiling of a large caliber nasogastric tube (Replogle tube) in the upper esophageal pouch. Chest and abdominal X-ray demonstrate intestinal gas, which indicates the presence of a distal tracheoesophageal fistula. Surgical repair via right thoracotomy or thoracoscopy is typically performed in the first 24–72 h of life in otherwise healthy neonates [2].

Low birth weight and associated cardiac anomalies are independent predictors of mortality in infants undergoing EA/TEF repair [3]. The survival rate after EA/TEF repair in less than 2 kg neonates with major cardiac anomalies is only 27% as compared to a survival rate of 100% in greater than 2 kg neonates without heart disease [4]. Due to the high incidence of associated

B. G. Goudra et al. (eds.), *Anesthesiology*, https://doi.org/10.1007/978-3-319-74766-8_39

Table 39.1 Summary of the plan of anesthesia with reasoning

Plan	Reasoning
The patient should be stabilized in the neonatal intensive care unit in a 30° head up position with continuous suctioning of the upper esophageal pouch	To minimize the risk of aspiration and the development of pneumonia
Preoperative intravenous fluid and antibiotic prophylaxis should be administered	To prevent dehydration, hypoglycemia and to reduce the risk of respiratory tract infection
Preoperative echocardiogram is required	Due to the high incidence of associated cardiac anomalies and to identify the side of the aortic arch
The key to airway management for TEF is the placement of the endotracheal tube with the tip lying below the fistula but above the carina	To provide adequate ventilation while minimizing gastric distention
Avoid awake intubation	Awake intubation in a vigorous baby, especially a preterm baby, can result in intraventricular hemorrhage
Standard management includes induction of general anesthesia with muscle relaxation and endotracheal intubation	The majority of TEFs are small and appear to be low risk for difficult ventilation, even if they are located below the tip of the ETT
Rigid bronchoscopy is also routinely performed in the same anesthetic prior to the onset of surgical repair of the TEF in many centers	To identify the location and size of the fistula, to characterize airway anatomy and to place a Fogarty catheter to occlude a large TEF
The end-tidal carbon dioxide ($EtCO_2$) tracing needs to be carefully monitored, together with prompt availability of a flexible bronchoscope, intubation equipment and suction	The small size of the ETT and working around the tracheal area during TEF repair pose additional risks of ETT dislocation and kinking and plugging of the ETT with mucus or blood
An arterial line is indicated in patients with congenital heart disease, prematurity, poor pulmonary compliance, and a large fistula with preoperative ventilatory compromise	These high risk patients require continuous monitoring of mean arterial pressure, frequent blood sampling and following of blood gas analysis
An arterial line is also indicated in patients undergoing thoracoscopic repair	During one lung ventilation, the $ETCO_2$ is falsely low and arterial blood gas is a better monitor of ventilation
The patient can be extubated immediately postoperatively in select patients. However, the majority of patients remain intubated in the neonatal intensive care unit after surgery	Extubation can be complicated by poor pain control, tracheomalacia, and vocal cord paralysis. If the patient requires emergency reintubation, there may be more disadvantages due to a traumatic intubation, leakage at the anastomotic site, or accidental removal of the trans-anastomotic feeding tube

cardiac anomalies (29%), preoperative echocardiogram is required in all patients undergoing TEF repair. Beside the heart structures, the position of the aortic arch also needs to be identified. In 2.5% of EA/TEF patients, the aortic arch is located on the right side. The left thoracotomy approach is much easier than the usual right thoracotomy incision in patients with a right sided aortic arch [1]. Preoperative lumbar ultrasound is beneficial in neonates with a sacral dimple due to the high incidence of vertebra anomalies if caudal catheter placement is planned for postoperative pain control.

Severe pneumonia or dependence on mechanical ventilations are no longer prognostic indicators of mortality in neonates with EA/TEF. The improvement in neonatal care including the use of maternal steroids and the availability of exogenous surfactant has significantly reduced the incidence of respiratory distress syndrome which complicated prematurity and aspiration pneumonia [5].

Anesthetic Management

Preoperative Preparation

The surgical repair of EA/TEF is urgent, but not emergent. Delay in surgical correction results in increased risk of aspiration and pneumonitis. The patient should be stabilized in the neonatal unit

and maintained in a 30° head up position with continuous suction of the upper esophageal pouch. Preoperative intravenous fluids should be administered to prevent dehydration and hypoglycemia. Prophylactic antibiotics are required to reduce the risk of respiratory tract infection.

Airway Management

The key to airway management for TEF is the placement of the endotracheal tube with the tip lying below the fistula but above the carina in order to provide adequate ventilation while minimizing gastric distension. A cuffed endotracheal tube (ETT) is preferable during the surgical repair. Not only does a cuffed ETT provide better tidal volumes with minimal leakage while facing increase in intrathoracic pressure during thoracotomy, the cuff can be used to occlude the TEF [6]. For the uncuffed ETT, use the ETT without a side hole and face the bevel of the ETT to the front of the trachea to minimize the chance of ventilation through the TEF in the back of the trachea [7].

Traditionally, the literature recommended awake intubation, the avoidance of positive pressure ventilation and the avoidance of muscle relaxant until the fistula was surgically controlled. There are many disadvantages of this approach. First, awake intubation in a vigorous baby, especially a preterm baby, can result in intraventricular hemorrhage. Second, adequate ventilation and good surgical conditions are difficult to achieve during open thoracotomy without muscle relaxant. Third, the majority of TEF are small and appear to be low risk for difficult ventilation, even if it is located below the tip of ETT [8]. Therefore, the current standard management is induction of general anesthesia with muscle relaxant and endotracheal intubation.

Rigid bronchoscopy is also routinely performed in the same anesthetic prior to surgical repair in many centers in order to locate and size the TEF. Rigid bronchoscopy can also identify other associated anomalies in the airway such as laryngomalacia, tracheomalacia, multiple fistulas, and external compression from a vascular ring [9]. When the fistula is large and lies just above the carina, maintaining proper position of the ETT is challenging. Adequate ventilation can be achieved by occlusion of this fistula using a Fogarty catheter with placement under rigid bronchoscopy visualization. With this approach, rigid bronchoscopy will be performed under general anesthesia while the patient is spontaneously ventilating. After identification of the location and structure of the fistula(s), the Fogarty catheter is placed via a rigid bronchoscope and inflated. Then, muscle relaxant can be given, followed by endotracheal intubation. The ETT is placed in the trachea, next to the Fogarty catheter. The proper placement of the ETT can also be checked using a flexible bronchoscope. After control of the TEF is obtained, the Fogarty balloon is deflated and removed while the ETT is still in place.

When a rigid bronchoscope or small fiber optic bronchoscope is not available, the conservative method is to place the ETT using direct laryngoscopy into the main stem bronchus. The ETT is then slowly pulled back until the breath sounds are heard bilaterally and the patient is well ventilated with minimal gastric distention. If conditions are not optimized, the patient may require one lung ventilation. The tip of the ETT must be on the opposite side of the surgical incision (lower lung in lateral decubitus position). The down side of bronchial intubation is bronchial edema resulting in prolong collapsed of the upper lobe [10]. After surgical repair, the ETT will be moved up into the trachea to achieve two lung ventilation.

General Considerations

The patient typically arrives in the operating room with intravenous fluids replacement, a suction in the upper esophageal pouch, and in the head up position. Aggressive bag mask ventilation should be avoided, since it leads to excessive gastric distention. After inhalational induction, if there is no plan for bronchoscopy, many pediatric anesthesiologists routinely paralyze the patient after successful gentle bag mask ventilation before intubation. Good intubating conditions

can be achieved with deep inhalational anesthesia, but thoracotomy without muscle relaxant is very challenging. With a small fistula, the tip of the ETT can be well above the fistula and still provide adequate ventilation. Either surgical ligation of the fistula or clamping of lower esophagus [11] will prevent further gastric insufflation. This approach avoids the need to re-tape and readjust the ETT position from a single lung to a tracheal intubation while the baby is under the drapes in the lateral decubitus position. Expansion of the lung may also be required to correct hypoxia during the procedure and at the end of the procedure to manage atelectasis.

Anesthesia for Gastrostomy

Preoperative gastrostomy is rarely performed due to the loss of the ability to ventilate and leakage of air through the open gastrostomy. A small TEF rarely affects the ability to ventilate. Rigid bronchoscopy and placement of a Fogarty catheter allow for occlusion of a large TEF prior to a definitive repair. In very rare circumstances, when the patient presents with respiratory compromise and requires emergency gastrostomy prior to definitive repair, spontaneous breathing should be maintained. After inhalational induction, a small dose of intravenous fentanyl and topical lidocaine on the vocal cords can be used to facilitate intubation. In premature infants with a severely dilated stomach, a large angiocatheter may also be placed into the abdomen to rapidly achieve a gastrostomy without general anesthesia. It is important to note, however, that adequate ventilation may not be achieved after stomach decompression due to air leakage via the gastrostomy. Temporary occlusion of the gastrostomy or retrograde occlusion via the gastrostomy site may be required to improve ventilation until the fistula is surgically controlled.

The most common indications of gastrostomy placement are as a part of a staged repair for feeding and to improve nutrition in very low birth weight neonates [12] while waiting for delayed primary repair, or in patients with long gap esophageal atresia.

Anesthesia for Open Thoracotomy Repair of TEF

It is essential to always obtain an echo cardiogram to identify the position and location of the aortic arch. Routinely, patients will undergo a right thoracotomy. If the patient has a right sided aortic arch, surgical approach will be much easier and safer via a left thoracotomy.

Besides routine care of a neonate undergoing general anesthesia (i.e. keeping the patient warm, placing standard ASA monitors, having two peripheral intravenous lines for fluid management), airway management and adequate ventilation are always challenging during TEF repair. The small size of the ETT and working around the trachea area pose the additional risks of ETT dislocation, or kinking and plugging of the ETT with mucus or blood. Ventilation during thoracotomy and surgical retraction is often inadequate. In addition to the standard ASA monitors, an arterial line is also required in high risk patients including those with congenital heart disease, prematurity, poor pulmonary compliance, and a large fistula with preoperative ventilatory compromise. An arterial line not only provides continuous monitoring of the mean arterial pressure, it also allows frequent blood sampling and following blood gas analysis. The end-tidal carbon dioxide ($EtCO_2$) tracing needs to be carefully monitored together with prompt availability of flexible bronchoscopy and intubation equipment as well as suction to interrogate the ETT if clogging or dislodgement is suspected.

After the TEF is ligated, the esophagus is then reanastomosed. The trans-anastomosis feeding tube is passed and becomes a critical tube. This feeding tube promotes nutrition by allowing enteral feeding and reducing the incidence of gastroesophageal reflux as compared to a gastrostomy tube [1]. A caudal catheter advanced to T6–T7 provides good postoperative analgesia and

avoids narcotics there by promoting early extubation [2]. In selective populations, the patient can be extubated immediately postoperatively. This results in less trauma from rubbing of the ETT against tracheal mucosa. However, extubation can be complicated by poor pain control, tracheomalacia, and vocal cord paralysis [5]. If the patient requires emergency reintubation, there may be greater disadvantages due to trauma, leakage at the anastomosis site, or accidental remove of the trans-anastomosis feeding tube.

Anesthesia for Thoracoscopic Repair of TEF

Surgeons are increasingly trying to repair TEFs via a thoracoscopic approach. The patients will be in the semi-prone position with 45° elevation of the right side with the right arm above the head. These patients receive the same intraoperative anesthesia management, however an arterial line is always required. Due to insufflation of carbon dioxide (CO_2) to collapse the right lung, ventilation during the thoracoscopic surgery is often compromised. In a case series by Krosnar et al. from Edinburg, all patients undergoing thoracoscopic repair desaturated to 84–91% and most of them required 100% oxygen during 5 mmHg of CO_2 insufflation [10]. During one lung ventilation, the $ETCO_2$ is falsely low and therefore serial arterial blood gas sampling is a better monitor of ventilation.

Postoperative Complications and Considerations

Major postoperative complications include sepsis, pneumothorax, unilateral or bilateral vocal cord paralysis, tracheomalacia, renal insufficiency, anastomosis leakage, esophageal stricture, gastroesophageal reflux and recurrent tracheoesophageal fistula [3]. Recurrent TEF occurs in 3–20% of patients after TEF repair depending on the TEF type and surgical technique. Endoscopic repair of recurrent TEF using a ventilating rigid tracheoscope simultaneous with flexible esophagoscope to de-epithelize and seal the fistula with fibrin glue, results in lower morbidity but higher rate of recurrence compared to open approaches. Ideal candidates for this technique are patients with a long, thin and small fistula located proximally where the cuffed endotracheal tube can be placed beyond the fistula site [13].

Long gap esophageal atresia is a gap wider than 3 cm or two vertebral bodies. Patients with long gap esophageal atresia can be a surgical challenge. Since a primary anastomosis is not possible, this group of patients require G-tube placement and delayed repair. Traction suture of the upper and lower esophageal pouch can be placed when a primary anastomosis is not feasible. This traction results in esophageal elongation which facilitates delayed primary repair. Besides esophageal lengthening, esophageal replacement using colonic interposition or gastric tube interposition has been proven effective. However, the risks of esophageal stricture, leakage, recurrent TEF, and esophageal mucosal outpouching increase with a complicated repair [14]. There is a strong association between gastroesophageal reflux, anastomotic tension and leakage of the anastomosis with esophageal anastomotic stricture [1].

References

1. Spitz L. Esophageal atresia. Lessons I have learned in a 40-year experience. J Pediatr Surg. 2006;41(10):1635–40.
2. Gayle JA, Gomez SL, Baluch A, Fox C, Lock S, Kaye A. Anesthetic considerations for the neonate with tracheoesophageal fistula. Middle East J Anaesthesiol. 2008;19(6):1241–54.
3. Diaz LK, Akpek EA, Dinavahi R, Andropoulos DB. Tracheoesophageal fistula and associated congenital heart disease: implications for anesthetic management and survival. Paediatr Anaesth. 2005;15(10):862–9.
4. Okamoto T, Takamizawa S, Arai H, Bitoh Y, Nakao M, Yokoi A, et al. Esophageal atresia: prognostic classification revisited. Surgery. 2009;145(6):675–81.
5. Broemling N, Campbell F. Anesthetic management of congenital tracheoesophageal fistula. Paediatr Anaesth. 2011;21(11):1092–9.
6. Greemberg L, Fisher A, Katz A. Novel use of neonatal cuffed tracheal tube to occlude tracheo-oesophageal fistula. Paediatr Anaesth. 1999;9(4):339–41.

7. Baraka A, Akel S, Haroun S, Yazigi A. One-lung ventilation of the newborn with tracheoesophageal fistula. Anesth Analg. 1988;67(2):189–91.
8. Andropoulos DB, Rowe RW, Betts JM. Anaesthetic and surgical airway management during tracheo-oesophageal fistula repair. Paediatr Anaesth. 1998;8(4):313–9.
9. Kane TD, Atri P, Potoka DA. Triple fistula: management of a double tracheoesophageal fistula with a third H-type proximal fistula. J Pediatr Surg. 2007;42(6):E1–3.
10. Krosnar S, Baxter A. Thoracoscopic repair of esophageal atresia with tracheoesophageal fistula: anesthetic and intensive care management of a series of eight neonates. Paediatr Anaesth. 2005;15(7):541–6.
11. Ni Y, Yao Y, Liang P. Simple strategy of anesthesia for the neonate with tracheoesophageal fistula: a case report. Int J Clin Exp Med. 2014;7(1):327–8.
12. Petrosyan M, Estrada J, Hunter C, Woo R, Stein J, Ford HR, et al. Esophageal atresia/tracheoesophageal fistula in very low-birth-weight neonates: improved outcomes with staged repair. J Pediatr Surg. 2009;44(12):2278–81.
13. Richter GT, Ryckman F, Brown RL, Rutter MJ. Endoscopic management of recurrent tracheoesophageal fistula. J Pediatr Surg. 2008;43(1):238–45.
14. Al-Shanafey S, Harvey J. Long gap esophageal atresia: an Australian experience. J Pediatr Surg. 2008;43(4):597–601.