Temporary Retrograde Occlusion of High-Flow Tracheo-Esophageal Fistula

Colin D. Gause, MD, Ian Glenn, MD, Michael Liu, MD, Federico G. Seifarth, MD

This report describes a temporary retrograde occlusion technique for control of a high-flow tracheo-esophageal fistula in a critically ill, premature infant born at 29 weeks' gestational age, with a diagnosis of type C (Gross) esophageal atresia and tetralogy of Fallot (TOF). This procedure is a useful bridging maneuver before definitive surgical correction for extremely low birth weight, unstable neonates with tracheo-esophageal fistula who are suffering from associated malformations.

Esophageal atresia (EA) with tracheo-esophageal fistula (TEF) represents a spectrum of foregut abnormalities characterized by incomplete esophageal organogenesis with or without associated abnormal communication between the trachea and esophagus, with an incidence of ~1 in 3500 births worldwide. Patients have additional congenital abnormalities in ~50% to 70% of cases. These abnormalities are predominantly cardiovascular (20%-39%), although genitourinary (14%-24%), gastrointestinal (14%-23%), musculoskeletal (17%-56%), and central nervous system (7%) abnormalities are also common. Cardiovascular anomalies contribute to the majority of morbidity and mortality, cited as the cause of death in 78% of cases.

Anatomic classification is most commonly described according to the Gross system, which classifies the disease into 6 categories designated A to F, the most common of which is type C: proximal EA with distal TEF (84% of cases). The Waterson classification stratifies these infants into survival categories A through C, based on birth weight and associated anomalies. This classification system was refined by Randolph et al. The authors used physiologic status as a guide in which those with stable cardiac and respiratory status underwent early repair, whereas unstable patients underwent delayed repair. Spitz et al later proposed a new classification system, designated I to IV, with emphasis on cardiac anomalies, because they represent the predominant predictor of survival.

Neonates with a large TEF are at risk for shunting excessive amounts of air into the gastrointestinal tract, and positive-pressure mechanical ventilation exacerbates this phenomenon, contributing to subsequent hypoventilation. Extremely premature neonates or patients with severe associated anomalies are often too unstable to undergo early TEF repair. Therefore, operative intervention to occlude the TEF before definitive repair may be necessary to allow adequate ventilation and prevention of gastrointestinal complications. Gastrostomy represents a potential option, but this method allows air to preferentially flow through the fistula and exit via gastrostomy at the expense of pulmonary ventilation.
occurrence is particularly problematic in neonates with poor lung compliance. Bronchoscopic placement of a Fogarty balloon to occlude the fistula,\textsuperscript{13,14} endotracheal (ET) tube placement distal to the fistula,\textsuperscript{12} esophageal banding,\textsuperscript{15} Nissen fundoplication,\textsuperscript{16} and gastric division\textsuperscript{17} are additional options to prevent diversion of airflow, with bronchoscopic occlusion used most commonly.

This case report describes the management of an unstable, premature, 810-g infant with a type C TEF and TOF (Waterson C, Spitz group III, Okamoto class IV), in which the high-flow fistula was managed with creation of a gastrostomy and fluoroscopically guided placement of a retrograde esophageal fistula occlusion balloon catheter, followed by staged repair of the TEF after stabilization of the patient’s status.

**CASE REPORT**

The patient is a premature female singleton, born at 29 weeks’ gestational age, with an ultrasonographic prenatal diagnosis of TOF. Before the second minute of life, the patient was intubated, and inhaled surfactant was administered for respiratory distress of prematurity. Attempts to insert a Replogle orogastric tube failed, because the tube could not be advanced >8 cm. Subsequent abdominal radiograph revealed diffuse gastric, small and large bowel gaseous distention, confirming the diagnosis of type C TEF. The Replogle tube was positioned in the distal esophageal pouch and kept to low continuous suction. Contrast radiography was not performed. Echocardiography confirmed TOF.

The patient remained stable for the first 3 days of life. However, despite delicate ventilator management, she developed increasing abdominal distention and needed progressive ventilator support, prompting urgent intervention. On day 4 she was taken to the operating room, where she underwent a 2.5-cm transverse mini-laparotomy, Stamm gastrostomy, and placement of a 10-Fr Mallinckrodt catheter. At the gastrostomy site, a 6-Fr Fogarty catheter was advanced under fluoroscopic guidance into the TEF (Figs 1 and 2). The balloon was cautiously inflated with 1 mL water-soluble radiopaque contrast until ventilator control indicated resolution of the air leak (Figs 3 and 4). The Fogarty catheter was secured to the abdominal wall. The Mallinckrodt was then temporarily placed to water seal to detect an air leak, of which there was none. Despite significant improvement of ventilator management, the patient was deemed too unstable to undergo repair at that time.

Over the course of her hospitalization, the Fogarty balloon was deflated every other day for ∼30 minutes to prevent accumulation and aspiration of esophageal secretions. Gastric tube feeds were begun. On day 25, she was taken to the operating room for ligation of the TEF. The fistula was temporarily occluded with a 6-Fr Fogarty catheter via the trachea, and the patient underwent uncomplicated thoracotomy and suture ligation of the fistula. The distal esophagus was tacked to the chest wall under mild tension and the proximal esophageal pouch remained intubated with a Replogle catheter. On day 145, she underwent complete repair of TOF, closure of her patent foramen ovale, and ligation of her patent ductus arteriosus. Repair of TEF was deferred for 6 weeks in accordance with the recommendations of our cardiac surgery colleagues. On day 197, the patient underwent definitive reconstruction of the esophagus via thoracoscopic esophageostomy. She tolerated the procedure well, and there were no complications.

The patient was extubated on postoperative day 5. Iohexol upper gastrointestinal series on postoperative day 7 revealed a patent anastomosis with contrast flow into the stomach, with no evidence of extravasation or anastomotic leak (Fig 5). She was started on breast milk by mouth on postoperative day 15, with supplemental gastrostomy feeds. She recovered well from her operations and continued to gain weight appropriately. The patient was discharged from the hospital into the care of her parents on day 221 of life, smiling and active. She was last seen in clinic at 14 months of age (8 months after repair). At that time, she was tolerating tube feeding at goal and taking puffed baby food and yogurt by mouth, with
adequate weight gain given her adjusted age.

DISCUSSION

Retrograde occlusion of a TEF via gastrostomy was first described by Hofmann in 1976.18 Hofmann placed the Fogarty balloon into the distal esophagus via open gastrotomy and used a silastic band around the cardia to perform a functional gastric division. Kadowaki et al19 similarly reported retrograde insertion of a Fogarty catheter into the distal esophagus in 4 low birth weight neonates, which were subsequently loosely ligated with sutures. Both techniques require large laparotomies to allow gastric and lower esophageal dissection. In 1985, Karl20 demonstrated that neither silastic band nor suture placement was necessary to achieve retrograde esophageal occlusion, performed in a premature 1270-g boy. In contrast to the aforementioned reports, we applied this technique in an extremely low birth weight, premature patient with TOF via mini-laparotomy. To the best of our knowledge, this case represents the first application of retrograde TEF occlusion in an extremely low birth weight (810 g) neonate with severe cardiac comorbidities.

Retrograde TEF occlusion offers advantages over the more common and invasive methods of occlusion. Unlike bronchoscopic fistula occlusion,13,14 this method avoids the resistance to airflow of a large-bore tube in relation to a neonatal trachea. Additionally, difficulties with the ET tube and associated adjustment will not interfere with balloon positioning. Compared with Nissen fundoplication, retrograde balloon occlusion is less invasive,16 and it avoids potential complications of the Nissen fundoplication, notably an excessively tight wrap interfering with subsequent feeding or an overly loose wrap resulting in persistent air leak. ET tube placement distal to the fistula12 may be possible only in a proximal fistula, because efforts to completely pass the fistula may result in ET obstruction at the carina or incidental mainstem bronchus intubation. Retrograde occlusion is temporary and does not alter gastric anatomy, thus not necessitating a reversal procedure17 or surgical band removal.15 It is important to note that although a gastrotomy is created to introduce the retrograde occlusion catheter, the same site is used for placement of an enteral feeding tube, which may be removed without an additional operation.

Retrograde occlusion is certainly not without risks. Inflating a balloon in the esophagus or TEF is accomplished under fluoroscopic, not visual guidance, and thus understandably raises concern about overdistention and compromise of the esophageal circulation. This concern was addressed intraoperatively via slow inflation of the balloon and instructions to our anesthesia colleagues to instill the minimum amount of fluid in the balloon to negate an air leak. Forceful placement of the Fogarty catheter also carries the risk of perforation of the esophagus or trachea.

There are important considerations about balloon placement. The balloon should be placed proximally enough to minimize formation of a cul-de-sac in which secretions could accumulate, with the risk for aspiration. However, too proximal balloon placement carries the risk of obstructing the airway. Although fluoroscopy was used to aid in balloon positioning, flexible bronchoscopy could be used to confirm initial balloon position and to assess airway compromise from proximal placement or overinflation.

CONCLUSION

Retrograde esophageal occlusion is a rarely used but highly useful method for controlling TEFs in premature infants.
method of pulmonary stabilization in a patient with type C TEF and respiratory failure. We have shown that it can be safely applied in a low birth weight premature neonate with associated TOF. Retrograde TEF occlusion with concurrent gastrostomy tube placement has the added benefit of allowing for feeding and decompressing of the stomach if necessary. This method of TEF occlusion allows stabilization of critically ill patients and treatment of associated defects before definitive TEF repair.

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ABBREVIATIONS

EA: esophageal atresia
ET: endotracheal
TEF: tracheo-esophageal fistula
TOF: tetralogy of Fallot

REFERENCES

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