OPEN ACCESS ATLAS OF OTOLARYNGOLOGY, HEAD & NECK OPERATIVE SURGERY

OPEN CERVICAL SURGERY FOR CONGENITAL H-TYPE TRACHEO-ESOPHAGEAL FISTULAE (TOF)

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Oesophageal atresia (OA) with or without a tracheoesophageal fistula (TOF) has an incidence of 1 in 2500 – 4500 live births.[1] Two distinct TOF classification systems are used, with the initial system by Vogt published in 1929, that was later modified by Gross in 1953 into five types of TOF *(Figure 1)*.[2,3]

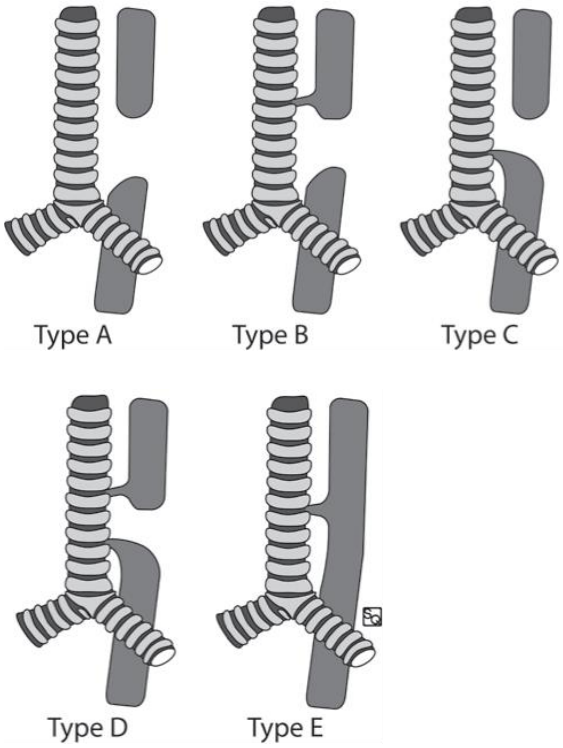


Figure 1: Gross TOF classification [4]

The most common TOF is Type C (86%) comprising of a proximal blind oesophageal pouch and distal TOF into the distal oesophagus *(Figure 1)*. Type E, better known as an H-type *(Figure 1)* fistula without OA, has an incidence of 4-7% of all TOFs.[4] H-type fistulae have occurs in 1 in 50,000 to 80,000 live births.[5]

Endoscopic procedures have been employed to obliterate TOFs and include using fibrin glue, electrocautery, or laser coagulation of the fistula tract.[6,7] Surgery remains the standard treatment with low recurrence rates.[4,8-10] An open cervical approach is recommended for TOFs located above the level of T2 [11] which accounts for majority of H-type TOFs.

Aetiology

The aetiology of TOF/OA is poorly understood. Although a TOF may occur in isolation, 50% are associated with congenital anomalies with cardiac anomalies being the most common. These associated anomalies are summarised in the genetic syndrome VACTERL (vertebral, anorectal, cardiac, tracheoesophageal, renal and limb abnormalities) *(Table 1)*. Other rare associated syndromes include Pierre-Robin, CHARGE or Di George with its chromosomal abnormality on 22q11 associated with oesophageal atresia.

Type	Incidence	Anomalies
Vertebral	17%	Scoliosis
Anal	12%	Imperforate anus, Duodenal atresia
Cardiac	20%	VSD, PDA, Tetralogy of Fallot, ASD
Renal	16%	Renal agenesis / dysplasia, hypospadias, polycystic
Limb	10%	Radial anomalies, polydactyly

Table 1: Congenital anomalies associated with TOF/OA [14]

Anatomy and Embryology

The trachea and oesophagus develop from a ventral diverticulum of the foregut. Proliferation of endodermal cells around week 3 of intrauterine life occurs over the diverti-

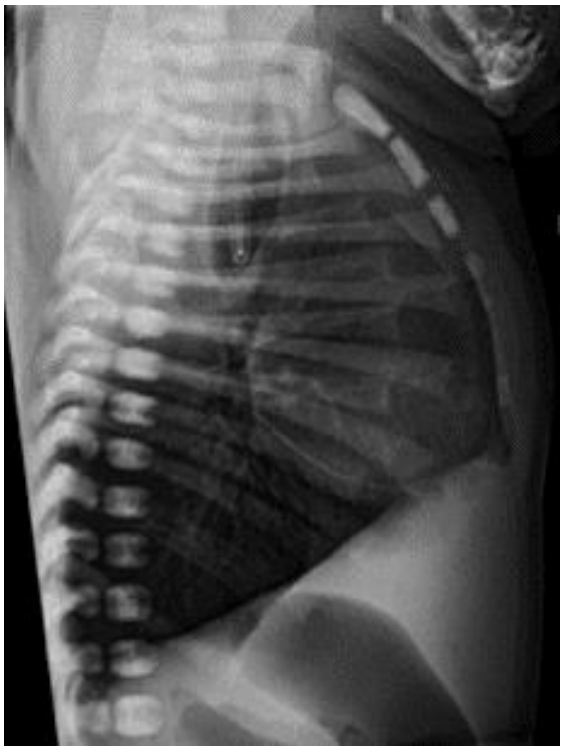
culum. The division of this cell mass forms the trachea and oesophagus. A TOF occurs during this period because of disruption to normal development. The primary mechanism of this disruption is unknown. However several key mutations including that of the *Sonic hedgehog* (Shh) gene identified in animal models result in failure of tracheoesophageal separation and fistula formation.[12]

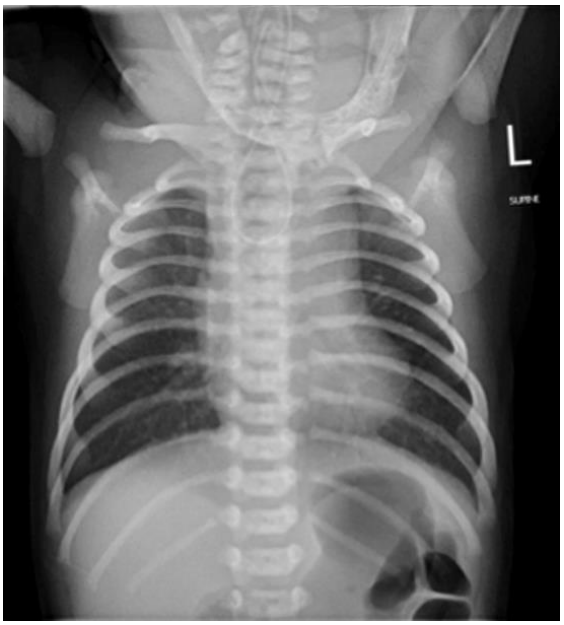
Diagnosis

Presentation and examination

TOFs are not easily diagnosed antenatally. Polyhydramnios identified on prenatal ultrasound may be secondary to oesophageal obstruction but is nonspecific and should not be used solely as a diagnostic marker.

The diagnosis of OA is made early following delivery and is confirmed by inability to pass a nasogastric tube into the stomach *(Figure 2)*.





Figures 2 a, b: Chest X-ray showing curled nasogastric tube at blind-ending oesophageal lumen and gastric bubble suspicion for TOF

The type of TOF is determined at microlaryngoscopy and bronchoscopy (MLB); however, subtle deformities and the level of a TOF can be missed *(Figure 3)*.

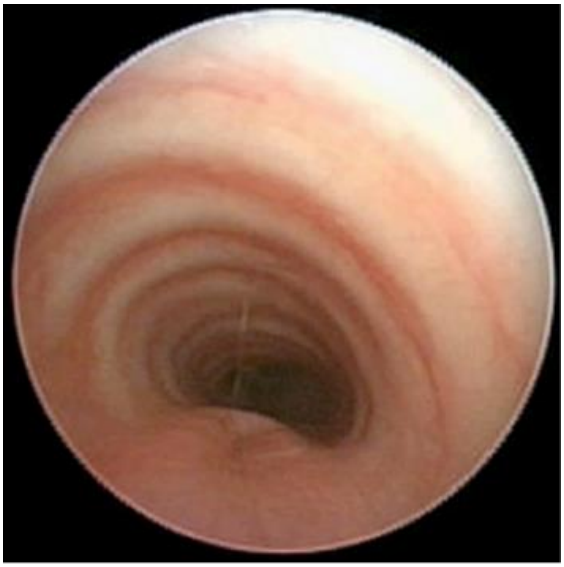


Figure 3: MLB with posterior tracheal wall TOF identified

H-type TOFs has more variable presentations. A delay or even misdiagnosis can occur as the oesophagus remains intact

without disruption. Common clinical symptoms include choking or cyanotic spells during feeding, abdominal distension leading to splinting during inhalation, or recurrent chest infections. These symptoms can be misdiagnosed as gastroesophageal reflux. A high index of suspicion for H-type TOF is important particularly with patients presenting with recurrent chest infections.

Investigations

Endoscopic evaluation including MLB and oesophagoscopy can identify the location, size, and number of TOFs, which are all important for surgical planning. Other associated laryngeal or tracheal abnormalities can be identified at the time of MLB.

Contrast radiographic studies (tube oesophagograms) are useful in premature and very small neonates where MLB is technically difficult. It avoids a general anaesthetic in neonates with complex underlying medical conditions. The neonate is placed supine or in a lateral position and contrast is injected into a nasogastric tube while slowly withdrawing it within the oesophagus. If there is a fistula, contrast will spill into the trachea. This is not routinely employed by the senior author *(Figure 4).*

Management

Immediate

A neonate presenting with difficulties with swallowing or respiratory distress with abdominal splinting following delivery as outlined above, should be transferred to a neonatal intensive care unit. A nasogastric tube (NGT) should be inserted, and its position confirmed on x-ray. Increasing abdominal distension with inhalation and a confirmed gastric position of a NGT should raise the suspicion of an H-type TOF *(Figure 5).*

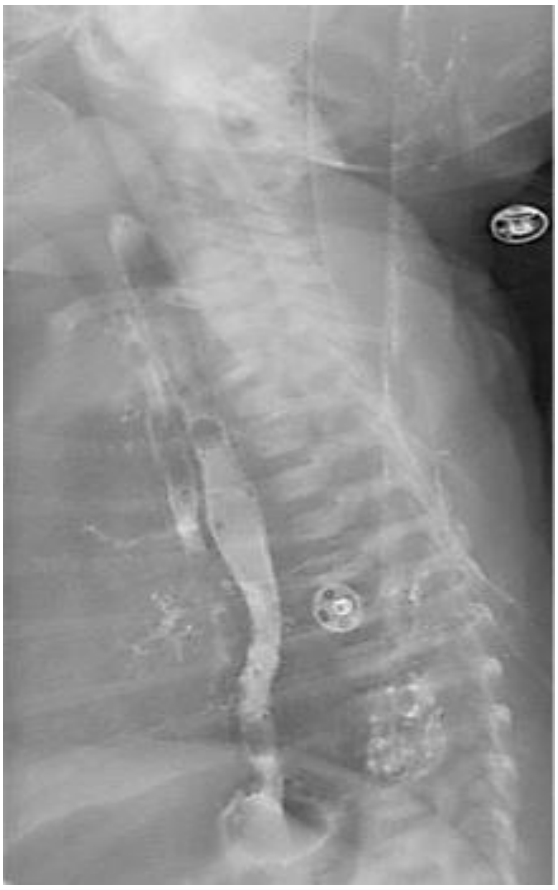


Figure 4: Contrast radiographic study showing communication between oesophagus and trachea

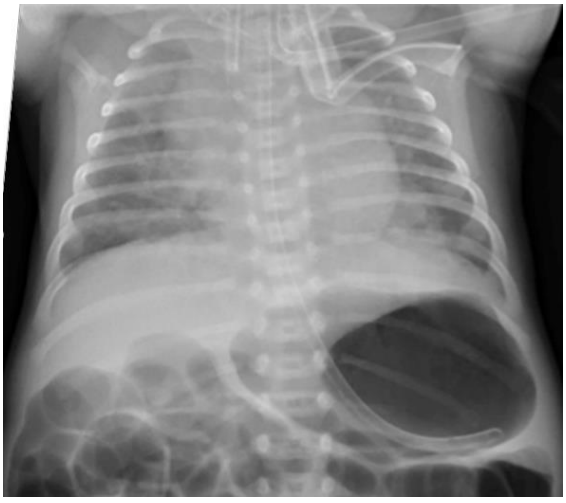


Figure 5: Chest X-ray showing the NGT in the stomach with significant gastric inflation from a suspected TOF

Aspiration of abdominal gas to alleviate abdominal splinting can be performed to

improve breathing. Neonatologists, paediatric ENTs, paediatric general surgeons, and paediatricians should be consulted.

Tracheoesophageal fistula identification

Initial management is directed at identifying the TOF, including the level, size, and number of fistulae. Using MLB, the TOF can be cannulated with a size 5/6 FG Foley catheter and then confirmed with oesophagoscopy *(Figure 6)*. TOFs located above T2 are amenable to open cervical surgery.

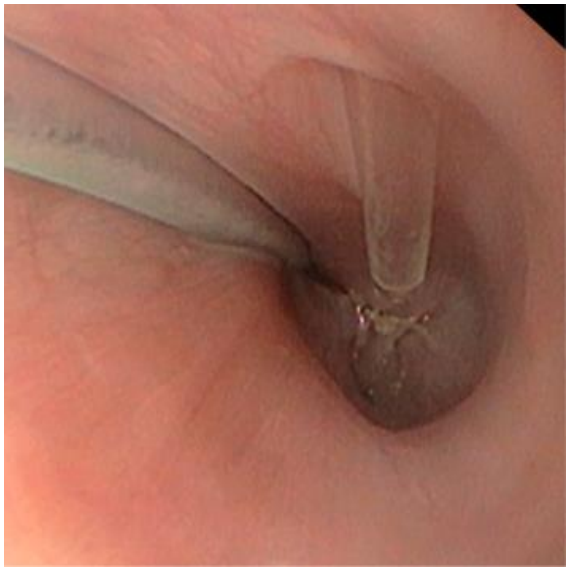


Figure 6: Oesophagoscopy identifying Foley catheter cannulated through the TOF from the trachea

Surgical techniques to close H-type TOF: open cervical approaches

Two open cervical surgical options can be used for the H-type TOF *i.e.*, ***extraluminal*** or ***transtracheal 'keyhole' techniques***. The open cervical approach is selected at the time of the MLB. Patients are intubated with an age-appropriate endotracheal tube (ETT) to confirm that the cuff is located distal to the TOF.

Extraluminal technique

- Following MLB, place the neonate or child in a supine position on a shoulder roll +/- head ring
- Administer a single dose of prophylactic antibiotics
- Identify anterior neck landmarks including thyroid cartilage, trachea, and sternal notch
- Mark a standard horizontal tracheostomy neck incision
- Infiltrate the subcutaneous tissues with local anaesthetic (1% lignocaine) and 1:100,000 adrenaline, not exceeding the maximum safe dose
- Prep the skin, and drape to protect the eyes, but to cover the mouth with the anterior neck and upper chest exposed
- Incise skin and subcutaneous tissue
- Raise subplatysmal flaps and retract the flaps with silk stay sutures
- Separate the strap muscles along the median raphe and retract them laterally
- Identify the trachea
- Divide the thyroid isthmus
- Create extraluminal dissection planes along the trachea up to the tracheoesophageal groove, staying on the tracheal adventitia
- Staying close to tracheal adventitia reduces the risk of injury to the recurrent laryngeal nerves
- Palpating a Foley catheter placed through the TOF at the time of the MLB can assist with identification of the TOF
- Having identified and circumferentially isolated the TOF, place a vessel loop around the TOF and withdraw the Foley catheter if previously placed through the TOF *(Figure 7)*
- Sharply divide the TOF
- Individually close the tracheal and oesophageal sides of the TOF

- Close the tracheal side with long lasting absorbable suture 4/0 PDS (Ethicon, Inc., Johnson and Johnson, New Jersey, US)
- Close the oesophageal defect using an inverting Connell suture technique with braided absorbable suture 4/0 Vicryl (Ethicon, Inc., Johnson and Johnson, New Jersey, US)

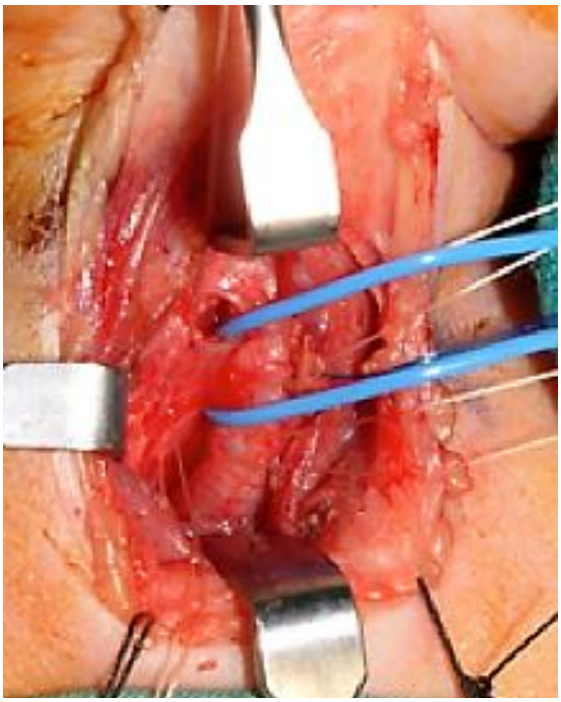


Figure 7: Extraluminal approach with vessel loop isolating the TOF and stay sutures securing the subplatysmal flaps [4]

- Reinforce the closure by interposing a local rotation muscle flap using one of the strap muscles, or a free graft of perichondrium from the *manubrium sterni*, secured with either 4/0 Vicryl and Tisseel glue (Baxter International Inc., Illinois, US)
- Do a layered closure of the neck with absorbable sutures over a Yates drain and apply a skin dressing

Transtracheal 'Keyhole' technique

The transtracheal technique reduces the risk of injuring the recurrent laryngeal nerves as it does not require circumferential extraluminal (tracheal) dissection. It permits a pouch-free closure of the trachea but requires an airtight seal and repair of the extended tracheotomy incision.

- Following MLB, place the neonate or child in a supine position on a shoulder roll +/- head ring
- Administer a single dose of prophylactic antibiotics
- Identify the anterior neck landmarks including thyroid cartilage, trachea, and sternal notch
- Mark a standard horizontal tracheostomy neck incision
- Infiltrate the subcutaneous tissues with local anaesthetic (1% lignocaine) and 1:100,000 adrenaline, not exceeding the maximum safe dose
- Prep the skin, and drape to protect the eyes, but to cover the mouth with the anterior neck and upper chest exposed
- Incise skin and subcutaneous tissue
- Raise subplatysmal flaps and retract the flaps with silk stay sutures
- Separate the strap muscles along the median raphe and retract them laterally
- Identify and expose the anterior trachea
- Divide the thyroid isthmus
- The number of tracheal rings exposed is dependent on TOF position and the tracheostomy site
- The level of the TOF has already been identified at MLB or is reconfirmed by flexible bronchoscopy via the ETT and withdrawing the ETT proximal to the TOF
- Place tracheal stay sutures at each side of the trachea

- Make a vertical extended transtracheal incision over the TOF position extending several rings inferiorly to incorporate a tracheostomy
- A temporary tracheostomy tube (reinforced / armoured ETT) is placed and secured to the anterior chest wall and the transoral ETT is removed
- The vertical tracheostomy incision is usually incorporated into the extended transtracheal incision, or can be a separate incision
- Identify the TOF on the posterior tracheal wall
- Cannulate the TOF with a Foley catheter transtracheally as a guide *(Figure 8)*

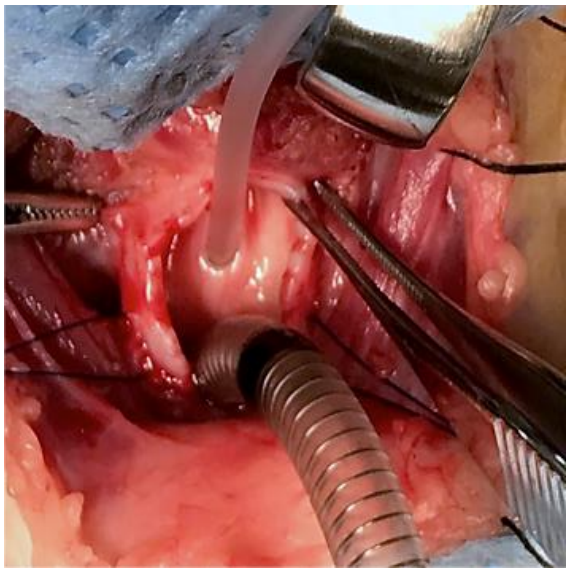


Figure 8: Transtracheal technique with extended vertical tracheostomy incision, temporary tracheostomy (reinforced ETT below TOF) and confirmation of TOF with Foley catheter cannulation

- Make an elliptical incision around the TOF in a vertical plane to isolate the TOF
- Dissect the TOF free of the trachealis muscle and retract it anteriorly with a stay suture *(Figure 9)*

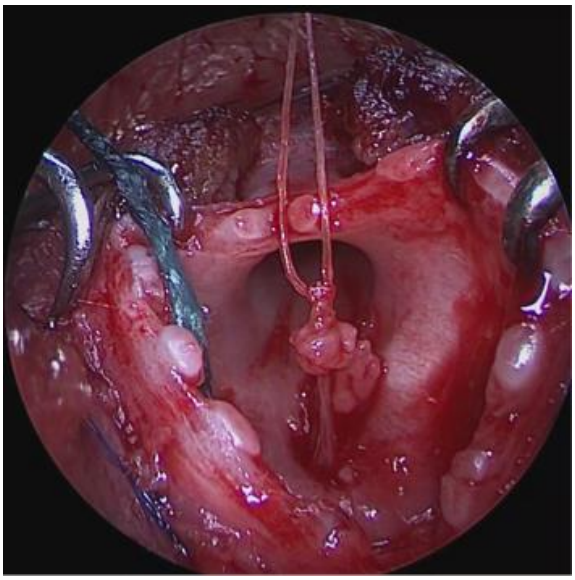


Figure 9: TOF isolated freely with elliptical incision from the posterior tracheal wall [4]

- Once isolated (remove Foley catheter if used as a guide), close the TOF with a Connell suture technique as for the extraluminal technique
- The repair can be reinforced through the posterior tracheal defect with periosteum harvested from the *manubrium sterni*
- Close the trachealis muscle with 5/0 Vicryl to create well-opposed muscle edges to reduce formation of a tracheal pouch *(Figure 10)*
- Intubate the child transnasally such that the cuff of the ETT is inflated below the level of the transtracheal incision, and mark the position of the ETT at the nasal aperture
- Remove the temporary tracheostomy tube
- Close the anterior tracheal incision with interrupted 5/0 PDS
- Tisseel glue can be applied over the tracheal repair
- Perform a Valsalva manoeuvre to confirm an airtight seal
- Do a layered closure of the neck with absorbable sutures over a Yates drain and apply a skin dressing

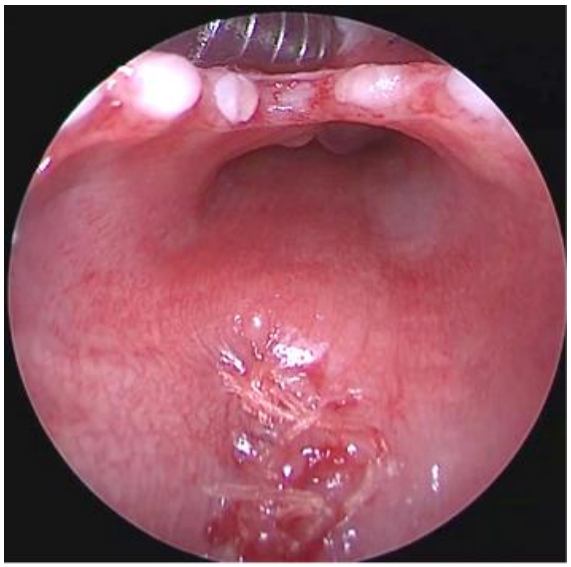


Figure 10: Intraluminal closure of the tracheal muscle [4]

Postoperative care

- Transfer the intubated patient to the neonatal or paediatric intensive care unit (NICU/ PICU)
- Extubate in NICU/PICU at 48-72hrs depending on clinical parameters
- Remove the Yates drain at 72hrs
- Continue IV antibiotics until the Yates drain is removed
- Request a contrast swallow study 1wk after surgery

Complications and long-term follow up

- ***Recurrent laryngeal nerve injury:*** Staying close to the tracheal adventitia with the extraluminal approach reduces the risk of injury
- ***Persistent TOF or recurrence:*** Reinforcing the closure with a rotation muscle flap and/or perichondrium reduces the small risk of failure and recurrence
- ***Surgical emphysema:*** Ensure an airtight tracheal closure
- ***Persistent, large, or recurrent TOF:*** These are difficult to manage surgically. Slide tracheoplasty has been used to treat these complex fistulae [13]

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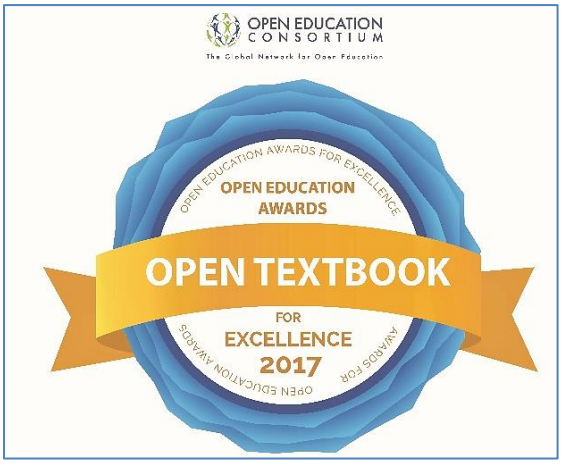

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