Acquired tracheo-oesophageal fistula in adults

Raman Diddee MB BS FRCA
Ian H Shaw BSc PhD MB BChir FRCA

The formation of an acquired tracheo-oesophageal fistula (TOF) is a rare but serious complication of malignancy and trauma. An established patent tract from the airway to the upper-gastrointestinal tract bypasses the normal protection offered by the laryngeal reflexes. As a consequence, this situation can present the anaesthetist with significant difficulties. The proximity of the oesophagus, trachea, upper mediastinal contents and large blood vessels can further complicate surgery and anaesthesia.1

Aetiology and pathophysiology

Over the last three decades the aetiology of acquired TOF has changed. Iatrogenic, malignant and traumatic causes have now superseded infection, formerly the predominant aetiology of acquired TOF.1-3 Presently, approximately 50% of acquired TOFs are secondary to mediastinal malignancy.1 Tumours arising from the oesophagus, trachea, lungs, larynx, thyroid and regional lymph glands have all been reported as prerequisites to the formation of an acquired TOF.4 The majority of acquired TOFs occur at the cervico-thoracic junction.

The non-malignant causes of acquired TOF are listed in Table 1. Of particular note is the increasing number of reports, in recent years, highlighting the ingestion of small camera batteries as a cause of acquired TOF in children.5

<table>
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<th>Table 1 Causes of non-malignant acquired TOF</th>
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<td>Blunt or penetrating trauma</td>
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<td>Granulomatous infection</td>
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<td>Previous surgery of trachea and oesophagus</td>
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<td>Corrosive fluid ingestion</td>
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<td>Poison and inhalation burns</td>
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<td>Tracheal tubes and intubation</td>
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<td>Percutaneous tracheostomy</td>
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Malignancy

Malignancy is the most common cause of acquired TOF. Of these, 77% are attributable to oesophageal tumours; 16% are secondary to pulmonary primaries.1-4 Oesophageal tumours pose the greatest risk with an incidence of 4.5%.4

The TOF forms as a result of necrosis and tissue breakdown. This is secondary to tumour enlargement and invasion but may be exacerbated by radiotherapy and chemotherapy. Untreated TOFs result in repeated airway soiling with rapid progressive pulmonary sepsis and death. The median survival from diagnosis is 1–6 weeks.4 This dismal prognosis warrants the surveillance of oesophageal tumours with bronchoscopy but even with limited treatment options postoperative mortality approaches 40%.3,4 Patients are often too unwell for surgery.

Acquired TOFs resulting from oesophageal malignancy are usually in the upper thoracic region where the oesophagus lies adjacent to the trachea.

Tracheal tube related

Of the non-malignant reports of acquired TOF in the literature, more than 75% are the result of endotracheal cuff-related trauma in patients subjected to prolonged mechanical ventilation. Secondary erosion of the tracheal and oesophageal walls occurs with a 0.3–3% incidence in mechanically ventilated patients.2

Tracheostomy does not appear to reduce the risk of developing an acquired TOF as a consequence of mechanical ventilation.

Reported mechanisms of injury include traumatic intubation, airway suctioning, and vascular compression of the tracheal wall resulting in ischaemia and subsequent ulceration. Tracheal wall ischaemia is a well-recognized complication secondary to prolonged endotracheal cuff over-inflation. The risk of developing an acquired TOF is further exacerbated if any of the predisposing factors listed in Table 2 are present.
The time taken for an acquired TOF to develop and become symptomatic is dependent on the precipitating cause and is illustrated in Table 3. More than one precipitating cause may be involved.

**Trauma**

Road-traffic accidents, resulting in chest wall crush injuries as a result of steering wheel impact, cause the majority of traumatic fistulae.\(^1\)\(^-\)\(^3\) Compression of the trachea and oesophagus between the sternum and the thoracic spine results in laceration and disruption of blood supply. This causes a delay in presentation of 3–10 days whilst necrosis develops.\(^1\) Although much less common, a more acute presentation can occur as a result of traumatic tracheal and oesophageal rupture. The TOF, which form mostly in the carinal area, have a reported operative mortality of 15%, predominately because of the rapid spread of mediastinal infection.\(^1\)

Ingestion of poisons and caustic substances causes local trauma and tissue necrosis. The most common paediatric aetiological factor in acquired TOF is the ingestion of button-type batteries.\(^5\)

**Granulomatous infection**

Although rare, acquired TOF can occur as a consequence of tuberculosis, HIV infection and mediastinitis. Infection as an aetiological factor in acquired TOF has declined in recent years.\(^1\)\(^-\)\(^3\)

**Surgical complication**

Post-oesophagectomy TOF occurs in 4% of patients, typically in the proximal and middle-third of the trachea.\(^4\) The majority are attributable to secondary ischaemia and devascularization of the trachea and main stem bronchi as a consequence of local surgical dissection. Predisposing factors include a transthoracic approach where access is restricted, dissection above the tracheal bifurcation, previous neo-adjuvant chemoradiotherapy and radical *en bloc* dissection.

Acquired TOFs are not infrequently associated with anastomotic incompetence where local inflammation and infection predispose fistulae formation. TOF in this category tend to be more distal, closer to the tracheal bifurcation and have a poorer prognosis.

**Pathological sequelae**

As a result of laryngeal bypass, spillage of oesophageal contents occurs into the trachea. Saliva, food and gastric juice contaminate the airways. This leads to congestion, infection, pneumonia, bronchial obstruction, atelectasis and respiratory distress. The severity of contamination depends on the width and length of the fistula as well as the posture of the patient. Spontaneous closure of non-malignant TOFs is exceptional.\(^1\)\(^-\)\(^4\)

**Clinical presentation**

An acquired TOF should be considered in any ventilated patient who has unexplained weight loss, recurrent chest infections and repeated failures to wean. Persistent tracheal soiling results in increased secretions on suctioning and subsequent pneumonia. Positive pressure ventilation can force gas through the patent fistula resulting in gastric dilatation and diaphragmatic splinting.

Symptoms in the non-ventilated patient are related to repeated tracheal soiling. ‘Ono’s sign’ refers to the uncontrolled coughing after swallowing, often worse with carbonated drinks. Other features which should raise suspicions of an acquired TOF are: history of trauma, malignancy or ingestion of caustic substances; chest pain; haemoptysis; shortness of breath; dysphagia; hoarseness; pyrexia of unknown origin; repeated respiratory tract infections; and pneumonia.

The patient with malignancy has the same symptoms in addition to those associated with the primary tumour. The lesion often becomes apparent after treatment of the primary tumour, particularly after chemotherapy or radiotherapy.\(^3\)\(^-\)\(^4\)

**Investigations**

A chest X-ray will demonstrate the effects of repeated soiling, fleecy basal infiltrates and the extent of the white-out revealing its severity. An over-inflated cuff, wider than the tracheal diameter, may suggest a cuff-related TOF. Barium swallow is possible if the patient is able to sit or stand. Contrast will demonstrate the defect in 70% of lesions.\(^2\)\(^-\)\(^4\) The site, width, length and direction of the TOF can be identified (Fig. 1).

Endoscopy is the best diagnostic method available for awake and asleep patients. Oesophagoscopy will enable the diagnosis of tumours and fistulae; biopsies may also be taken. Small TOFs may be missed in the folds of the oesophagus. Flexible or rigid
bronchoscopy identifies the TOF orifice better on the smooth posterior membranous wall and facilitates tissue biopsies. The instillation of methylene blue into the oesophagus can help pinpoint the TOF. Bronchoscopy also allows broncho–alveolar lavage, enabling targeted antibiotic therapy and airway clearance; both of these have been shown to improve outcome. Accurate identification of the site of the TOF is central to successful definitive management.

Preoperative management

The principles of preoperative management of an acquired TOF are to minimize further aspiration, prevent and treat pulmonary infections, provide supportive therapy until definitive surgery can be performed and ensure that the patient is as fit as possible for the surgical procedure. In particular, cardio-respiratory function should be optimized.

To prevent tracheal soiling, active methods of protection must be used. A new endotracheal tube (ETT) or tracheostomy tube should be passed with the cuff lying beyond the lesion if possible. The bed head should be elevated to prevent further passive regurgitation and aggressive pulmonary toileting should be used. Nasogastric tubes should be removed and replaced with a draining gastrostomy tube to reduce reflux in addition to acid suppression therapy.

In persistent soiling and carinal TOF, there is a case for oesophageal diversion. A gastrostomy is placed for feeding and the oesophagus is divided at the gastro-oesophageal junction. This carries a considerably higher rate of morbidity and mortality. Oesophageal diversion is useful in symptom palliation for malignant cases.

For the ventilated patient, supportive therapy is maintained and active measures are made to wean from the ventilator before surgery. This is critical for a successful outcome after surgical repair as patients should be extubated as soon as possible. It has been shown that postoperative positive pressure ventilation is associated with an increased incidence of anastomotic breakdown and stenosis, especially after tracheal resection. Therefore, it is prudent to wait until the patient has been weaned off the ventilator.

Feeding should continue and, as nutrition, infection and general status improve, ventilatory weaning can be commenced. The aim is for a single stage repair with rapid extubation avoiding postoperative ventilation.

Operative management

Surgery

For proximal TOFs the surgical approach is via an anterior or low cervical incision. Small lesions are repaired in two layers over a nasogastric tube. A muscle flap from the neck or intercostals is used to buttress the closure and increase success. TOFs associated with large circumferential lesions, such as may occur after pressure necrosis from an ETT cuff, may require tracheal resection, a description of which is beyond the scope of this article.

Anaesthesia

Anaesthetic management follows the same principles as induction for broncho-pulmonary fistula where prevention of tracheal soiling and provision of surgical access can be problematic. The surgeon and the anaesthetist are competing for airway access. Pre-medication with acid suppression therapy reduces gastric acidity and fluid volume. The site and size of the lesion must be carefully noted as this may dictate the anaesthetic approach. Only experienced thoracic and upper-gastrointestinal anaesthetists should carry out these cases.

Along with standard monitoring, direct arterial blood pressure measurement is useful, particularly as surgical manipulation of the neck can result in cardiovascular instability. The routine placement of a central venous catheter aids in fluid management.

Rapid isolation of the TOF is the key to successful anaesthetic management. Fortunately, most TOFs presenting for surgical
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repair are in the upper two-thirds of the trachea. If the tip of the ETT lies above the TOF, gastric dilatation and aspiration can occur. Cricoid pressure will be ineffective. Intubating the lumen of a large TOF with failure to ventilate is a major concern.

For confirmed high proximal tracheal TOFs, full preoxygenation followed by i.v. induction and neuromuscular paralysis with the patient in the semi-recumbent position may suffice. Succinylcholine or rocuronium will facilitate prompt intubation avoiding the need to apply positive pressure ventilation and the risk of gastric dilatation. If succinylcholine is used, pre-curarization is advisable to prevent muscle fasciculations increasing the risk of tracheal soiling and aspiration. The ETT can be placed under direct vision with a fibreoptic bronchoscope until the ETT cuff lies below the TOF. An awake fibreoptic intubation with the patient maintaining spontaneous breathing until the TOF is isolated is an alternative approach. A nasogastric tube, sited before induction, and left on free drainage, will help avoid gastric dilatation.

Carinal and bronchial TOF are rare but can present the anaesthetist with major difficulties. The site of the lesion means that protection of both lungs with a standard tracheal tube will be impossible. The choice must be made to isolate only one lung or to use an alternative. Passing a fibreoptic scope under the same conditions (as described above) will allow visualization of the lesion and allow the passage of the scope past the lesion into the unaffected bronchus. An endobronchial or standard tube may now be passed over the scope and single lung ventilation can occur. A double lumen tube may also be used to allow suctioning to the tracheal tube during surgery but these can be awkward, although not impossible, to pass over a fibreoptic scope. In paediatric cases, the use of cardiopulmonary bypass makes repair easier.

Once the TOF is isolated, ventilation can continue without fear of soiling and gastric dilatation. Anaesthesia can be maintained with a volatile anaesthetic agent or total i.v. anaesthesia and muscle relaxation. There are reports in the literature of TOFs being repaired whilst the patient breathes spontaneously, although assisted ventilation was often necessary. Surgery can last up to 4 h. After a middle or high TOF repair has been completed the ETT is withdrawn proximally and a leak test is performed. Following this normal anaesthesia continues. Ventilatory modes should be adopted that avoid unnecessarily stressing the surgical repair. Pressure-controlled and high frequency ventilation have all been reported but the literature fails to preferentially support any specific ventilatory mode.

Immediate extubation is the goal, so avoiding any postoperative ventilation. Extubation is achieved with careful use of opioids and neuromuscular blocking agents. Level 2 care is appropriate and facilitates careful monitoring of any respiratory complications. Analgesia will depend on the site of surgery. For the majority with proximal fistulae involving cervical incisions, local infiltration and systemic analgesia is usually sufficient. The less common distal repairs may be amenable to intrapleural analgesia, paravertebral blockade or even a thoracic epidural, as well as systemic analgesia.

**Oesophageal stenting**

TOFs secondary to a malignancy are often inoperable. In order to isolate the TOF and prevent pulmonary aspiration, oesophageal stenting can be undertaken. The stent is usually placed endoscopically under conscious sedation, thus negating the need for a general anaesthetic. Early stents were rigid plastic tubes that required aggressive oesophageal dilatation with the risk of tearing the TOF. The stents themselves were prone to migration, obstruction and were painful. Modern self-expanding metallic stents require no dilatation and are better tolerated. Tracheal stents have also been used but are beyond the scope of this article.

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**References**


Please see multiple choice questions 11–13.