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MISCELLANEOUS

Criticality in tailoring the treatment for tracheoesophageal fistulas in children

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Abstract

Objective Tracheo-oesophageal fistula (TOF) is a rare pathology. It can be congenital and concurrent with other congenital anomalies such as oesophageal atresia, laryngeal and tracheal agenesis, or it can be acquired. The purpose of this study was to analyse various management strategies and their outcomes in infants and children with TOF and identify potential areas for standardisation of the fistula repair procedures.

Methods At a single institution, a retrospective analysis of infants and children with congenital or acquired TOF between 2013 and 2019 was performed. Thirteen patients were identified. Data collection included: patient demography, associated congenital anomalies, details of fistula at the time of endoscopy, surgical approach and intra-operative findings, need for additional intervention(s), and outcomes.

Results Thirteen patients underwent endoscopic or open surgeries for correction of TOF. The TOF was congenital in ten patients and acquired in three patients. Eight patients had associated aero-digestive comorbidities, and six patients had systemic comorbidities. Three patients underwent endoscopic procedures and nine patients underwent an open TOF repair. One patient had tracheal agenesis and was not offered any treatment. Two patients required multiple endoscopic interventions for recurrent TOFs. Among four patients with prior tracheostomy, three were decannulated and one awaits decannulation.

Conclusıon

Appropriate case selection and surgical ergonomics are essential for patients with TOF to avoid recurrences. Preoperative endoscopy to obtain precise details regarding associated laryngotracheal lesions and demographics of the fistula is crucial.

Keywords Tracheoesophageal fistula · Aero-digestive anomalies

Introduction

Tracheo-oesophageal fistula (TOF) is a rare, congenital or acquired anomaly that is challenging for surgical treatment and carries significant morbidity and mortality if left unrepaired. It can be present independently or combined with various congenital anomalies. The time of onset of symptoms after birth varies. The overall prevalence of TOF is 2.82 per 10,000 live births and stillbirths [1]. It is manifested in childhood as recurrent pneumonias, and despite restricting oral intake, the patients have salivary and gastric reflux bronchoaspiration. This causes recurrent long duration hospitalisations and severe growth retardation.

Congenital TOF can co-exist with oesophageal atresia (OA), and the worldwide incidence of OA/TOF is approximately 1 in 2500–4500 live births [2]. OA/TOF is seen with laryngeal or laryngotracheo-oesophageal clefts (LTOC) in 20–37% of the cases [3]. The prevalence of laryngeal agenesis (LA) is unknown while tracheal agenesis (TA) occurs in 1 in 50,000–100,000 live births [4]. Prenatal ultrasonography and magnetic resonance imaging are essential in the diagnosis of LA and TA [5]. TA is incompatible with life, but there have been isolated cases of successful rescue procedures although with a poor prognosis [3]. The fistula associated with OA is distally located in 85% of cases and can be

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proximal or distal in cases of LA and TA [4]. Various genetic disorders (Feingold syndrome) can present with isolated congenital TOF [1] and severe tracheobronchomalacia. In all patients, it is important to rule out associated chromosomal anomalies or syndromes such as VACTERL association, trisomy 18, trisomy 21, trisomy 13, and CHARGE syndrome.

Acquired TOF may develop because of cuff-related injuries of the endotracheal tube, tracheostomy, ingestion of foreign bodies (alkaline batteries) or caustic injuries. Residual TOF is a term used for opening of the lower end of a LTOC repair or opening following OA with TOF closure (seen in 15% of the cases) due to unfavourable cicatrisation [6, 7]. Following endoscopic LTOC repairs, either residual fistula can occur due to technical errors in the surgical closure or secondary to a pre-existing tracheostomy with the cannula traumatising the distal cleft repair [5, 7].

Different approaches for managing TOF have been described. Endoscopic techniques [8–10] are used for small defects, and standard cervico-thoracic approaches are recommended for larger and longer fistulas [8, 9, 11]. This study highlights a single institutions' experience in managing diverse etiology TOFs and proposes treatment approaches for patients with different clinical presentations.

Materials and methods

After an internal review board clearance, we retrospectively collected data on all consecutive pediatric patients treated for TOF at the Lausanne University Hospital, Switzerland, between 2013 and 2019. The hospital electronic medical records with specific diagnosis codes were used to obtain the data. Two patients were treated in their home countries under supervision of the senior author (KS). All other patients were referrals from other tertiary Swiss or international centres sent exclusively for the treatment of the TOF and the underlying airway pathology. Patients with primary repair of EA and TOF without recurrence were excluded from this study.

To establish diagnosis and characterize the nature of the disease, all patients underwent flexible and rigid endoscopy (laryngotracheobronchoscopy and oesophagogastroscopy) and radiological studies. After diagnosing the TOF, the fistula site and dimensions were meticulously recorded using rigid 0° and angled telescopes (Storz, Germany). If a tracheostomy was present, the cannula was removed temporarily to document the oeso-tracheal fistulous tract. In cases of small suspicious TOF and to confirm the results after surgical repair, dilute 1% methylene blue dye test (Fig. 1) was performed by passing the coloured solution through a naso-pharyngo-esophageal tube, which was withdrawn gradually avoiding a spill over of the solution into the trachea. The presence of fistula or its repair integrity was confirmed by simultaneously passing a long thin 0° endoscope into the airway.

Patient demography, TOF etiology and its demography and associated aero-digestive pathologies were noted from the medical charts. Previous surgical procedures, intra-operative findings, outcomes (feeding, respiration) and complications were noted. All patients had at least 12 months follow-up and were regularly evaluated by the referring doctors. The information regarding their current quality of life, feeding and breathing was obtained by sending out electronic mails to the referring clinicians and parents who used the pediatric health-related quality of life (HR-QOL) instrument (12) to report the outcomes at 1 and 12 months after the surgery.

Microsoft Excel (Redmond, WA) was used to record and analyse the patients' data.

Results

Patients

The patient demography, TOF etiology and associated comorbidities are mentioned in Tables 1 and 2. Thirteen children (age 0–17 years) were identified. Ten patients had TOF secondary to a congenital background and three were acquired. The congenital group included five recurrences after EA and TOF repair, two after LTO cleft repair, one with Feingold syndrome and one each with laryngeal and tracheal agenesis. The three acquired TOFs included two

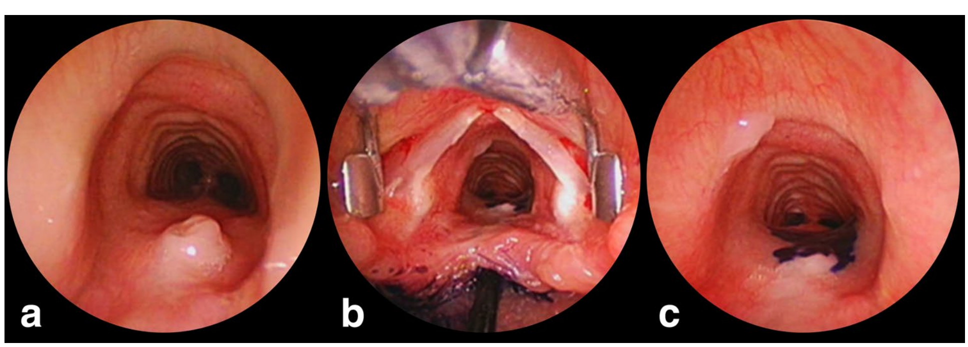

Fig. 1 The methylene blue dye test. **a** Tracheoesophageal fistula seen in the mid-trachea. **b** Methylene blue dye is passed into a naso-pharyngo-esophageal catheter. **c** Passing of the dye through the fistula tract is identified in the trachea

Table 1 Demography of TEF patients with congenital background

Patient	Sex	Age at presentation (days/ months/ years)	Respiratory distress	Feeding	Tracheotomy	Past surgery and associated aerodigestive pathology	Systemic comorbidities
1	M	5 months	Mild	PEG	No	Operated Type III B EARecurrent TEF Type 1 laryngeal cleft	Pulmonary artery stenosis, undescended testes, ventricular septal defect
2	F	4 years	Mild-moderate	PEG	No	TBM	Feingold syndrome
3	M	At birth	Severe	Died at birth	No	Tracheal agenesis EA	Atrioventricular septal defect
4	M	13 months	Mild	Oral (occasional aspiration with liquids)	Yes	Laryngeal agenesis Delivery by EXIT and tracheostomy	None
5	M	4 years	Moderate	PEG	No	Operated Type 3 LTEC Residual TEF Tracheomalacia	GER
6	M	4 days	Severe	PEG	No	Operated Type III B EA Severe TBM	GER
7	M	17 years	None	Oral (aspiration with liquids)	No	Type III B EA (operated on day 4 of life)	None
8	M	22 months	Moderate	PEG	yes	Operated Type 3 LTEC Long cicatricial tracheal stenosis	BPD
9	M	7 years	None	PEG	Yes	Operated Type III B EA (day 3) Grade I SGS Severe suprastomal collapse	None
10	F	4 years	None	PEG	No	Type III B EA Recurrent TEF (4 prior endoscopic attempts)	None

EA esophageal atresia, *EXIT* ex utero intrapartum, *TBM* tracheobronchomalacia, *LTEC* laryngotracheoesophageal cleft, *GER* gastroesophageal reflux, *BPD* bronchopulmonary dysplasia, *SGS* subglottic stenosis

Table2 Demography of patients with acquired TEF

Patient	Gender	Age at presentation (Years)	Etiology	Respiratory distress	Feeding	Tracheotomy	Associated aerodigestive pathology	Systemic comorbidities
11	F	2	Tracheotomy-induced	Mild	PEG + oral occassional aspiration with liquids	Yes	Intubation-induced severe transglottic stenosis	Prematurity
12	M	3	Battery ingestion	No	PEG	No	None	None
13	M	6	Battery ingestion	No	PEG	No	None	None

secondary to battery ingestion, and one was tracheostomy induced. All patients presented with symptoms of aspiration and multiple past hospitalisations for recurrent pneumonias. One infant with severe tracheomalacia had several episodes of dying spells at home that required emergency treatment on multiple occasions. The study included a patient with tracheal agenesis and congenital TOF, but was not offered any active management. The parents were informed of the bad prognosis and the child succumbed to his severe anomalies within minutes after birth.

TOF patients having a congenital etiology background

The timing when treatment for TOF was given varied between 1st day of life and 17 years. Seven out of ten patients were exclusively PEG fed, 1 (17 years) suffered from chronic aspiration and received no active treatment for this fistula until his referral to our clinic, 1 (13 m) with laryngeal atresia had a tracheostomy during the EXIT (ex-utero intra partum) procedure and was exclusively breast fed and had multiple hospital visits for aspiration pneumonia. Seven patients had significant airway lesions that needed concomitant treatment. These airway lesions included: severe tracheomalacia ($n=3$), laryngeal agenesis ($n=1$), type 1 laryngeal cleft ($n=1$), subglottic stenosis with suprastomal collapse ($n=1$) and tracheal stenosis ($n=1$). Six patients had systemic and two had syndromic comorbidities. One of these two patients had Feingold syndrome (with microcephaly, limb malformations, and learning disability) and the other had pulmonary artery stenosis with ventricular septal defect.

Except the deceased patient with tracheal agenesis, all patients underwent fistula repair surgery. The details of TOF correction and concomitant management of the airway lesions are mentioned in Table 3. Three patients had endoscopic treatment for their fistula, of which one had concomitant endoscopic closure of his laryngeal cleft. The fistula was exposed using rigid bronchoscope with the bevel facing towards the TOF. A gentle upward tug of the bronchoscope opened up the tract that was scarified using 33% trichloroacetic acid on a cytology brush and plugged using bioglue (Tisseel[R]). The child with Feingold syndrome (Fig. 2a–c) had multiple fistulas adjacent to each other and required three additional interventions before becoming asymptomatic. During his endoscopic repair, an extensive peri-fistulous chemical cauterisation was done to induce cicatricial fibrosis in the membranous trachea and thus treat the posterior to anterior floppy airway.

Open TOF repair interventions included: transtracheal closure of the fistula ($n=1$); tracheal resection and anastomosis ($n=4$); extended cricotracheal resection ECTR ($n=1$); and cervical slide tracheoplasty CST ($n=1$). Concomitant airway corrective surgeries included: rigidification of segmental tracheal malacic airway using multiple polydiaxanone microplates (Rapidsorb-Synthes[R]) ($n=1$); and posterior tracheopexy ($n=1$).

A temporary distal tracheostomy was performed during the transtracheal fistula closure and the tracheal and oesophageal mucosae separated using microinstruments (Fig. 3a–c). The oeso-tracheal mucosae were closed with a free patch of sternal periosteum interposed between the two layers.

The child with laryngeal atresia underwent an extended CTR (ECTR). The oesophagus was closed vertically in two layers (mucosa–submucosa and trachealis). Additional reinforcement was obtained with the rib cartilage that was used to expand the airway (as part of the ECTR) and further covered by the tracheal mucosal flap during glotto-tracheal anastomosis.

During tracheal resection and anastomosis (TRA), care was taken to avoid juxtaposing the oesophageal and tracheal sutures. The anterior oesophageal closure was vertical, as against the horizontal posterior tracheal repair. Sternal periosteum was used to interpose between the tracheal and oesophageal anastomosis (Fig. 3d, e).

One patient had tracheostomy-induced long tracheal stenosis involving six rings and we felt a TRA would be of risk in causing an airway dehiscence in him. Therefore, we performed a cervical slide tracheoplasty (CST) to close the TOF and concomitantly treat the tracheal stenosis.

One child underwent posterior tracheopexy during the primary repair of his EA. This was done intraoperatively under endoscopic guidance with the child breathing spontaneously, a flexible bronchoscope (3.1 mm Olympus) was passed through the endotracheal tube and withdrawn gradually up to the glottis. The floppy posterior tracheal wall was fixed with the prevertebral fascia and anterior spinous processes using resorbable sutures.

Check endoscopy was done in all patients after 7–10 days. It included a dynamic airway examination to check for airway stability and the methylene blue dye test to confirm fistula closure.

Patients with TOF secondary to an acquired etiology

Three patients (2, 3 and 6 years) were included in this group and were exclusively PEG fed. One patient had severe transglottic stenosis (TGS) and two patients had TOF secondary to battery ingestion. The TOF demographics, associated airway lesions and surgery done are mentioned in Table 3.

The child with TGS received an extended CTR. The two patients with battery ingestion had only aspiration symptoms and underwent single stage tracheal resection and anastomosis. The sizes of their fistulas were larger than seen during the pre-operative endoscopy and underwent 3 and 5 tracheal ring excision, respectively.

Postoperative outcomes

In our series of 13 patients, surgical repair (three endoscopic and nine open) was performed on 12 patients except the one deceased patient with tracheal agenesis. None of the patients

Table 3 TEF demography and treatment details

Patient	Fistula demography (site and size)	Treatment for TEF	Follow up duration (months)[b]	Airway pathology that was treated concomitantly	Current feeding status	Current respiratory complaints
Congenital						
1	Size: 2 mm Site: 3 tracheal rings proximal to carina	Endoscopic	46	Type 1 laryngeal cleft (suspension microlaryngoscopy, CO2 laser and endoscopic suturing)	Oral	None
2	multiple adjacent TEFs Size: 2 mm/ 2 mm / 1 mm Site: 2 tracheal rings proximal to carina/ 4–5 tracheal rings proximal to carina/ 5–6 tracheal rings proximal to carina	Endoscopic (× 3 attempts)	41	TBM	Oral	None
3	Size: 3 mm Site: proximal esophagus	Died	–	Patient with tracheal agenesis and offered no treatment	–	–
4	Size: 2 mm Site: at the level of 1st–2nd tracheal rings	Extended CTR (excision of 2 tracheal rings) and primary closure of the TEF	21	Laryngeal atresia	Oral	Awaiting decannulation (tolerates full cap)
5	Size: 3 mm Site: at the level of 3rd tracheal ring	Transtracheal closure with rigidification of the trachea with polydiaxanone bioresorbable microplates	29	TBM	Oral	None
6	Size: 2 mm Site: 2 tracheal rings proximal to carina	Primary posterior tracheopexy during EA correction (right thoracotomy)	26	Severe TBM	Oral	None
7	Size: 10 mm Site: 8 tracheal rings proximal to carina	TRA (excision of 3 rings)	27	None	Oral	None
8	Size: 3 mm Site: at the level of 2nd tracheal ring	Cervical ST with a distal mini tracheostomy	11	Tracheostomy induced long tracheal stenosis (6 rings)	Oral	Decannulated after ssACCG and external tracheal rigidification with polydiaxanone bioresorbable microplates
9	Size: 10 mm Site: at the level of 2nd tracheal ring	ssTRA (5 rings excised) + anterior cricoid split	28	Severe suprastomal collapse Grade I SGS	Oral	Decannulated
10	Size: 3 mm Site: at the level of 2nd tracheal ring	Endoscopic	25	None	Oral	None

Table 3 (continued)

Patient	Fistula demography (site and size)	Treatment for TEF	Follow up duration (months)[b]	Airway pathology that was treated concomitantly	Current feeding status	Current respiratory complaints
Acquired						
11	Size: 3 mm Site: Between 3rd-4th tracheal rings (facing opposite to the suprastomal collapse)	Extended CTR (excision of 5 rings)	34	Severe transglottic stenosis	Oral	Decannulated
12	Size: 8 mm Site: at the level of 4th tracheal ring	TRA (excision of 3 rings)	26	None	Oral	None
13	Size: 12 mm Site: at the level of 3rd tracheal ring	TRA (excision of 5 rings)	31	None	Oral	None

TBM tracheobronchomalacia, *CTR* cricotracheal resection and anastomosis, *TRA* tracheal resection and anastomosis, *ssTRA* single stage TRA with concomitant resection of the tracheostoma, *ST* slide tracheoplasty, *ssACCG* single stage anterior costal cartilage graft, *EA* esophageal atresia, *SGS* subglottic stenosis

[b]From time of surgical intervention

who underwent open surgeries had any complications and did not receive any additional intervention. One out of three patients who underwent an endoscopic repair required three additional endoscopic procedures for complete healing of the fistula. None of the patients who received endoscopic treatment needed an open repair. After complete closure of their fistula, all patients resumed oral feeds and had progressive normalisation of the food texture. Currently, all patients take oral solid and liquid feeds and have no bronchoaspiration.

Patients with airway malacia had progressive improvement in their breathing and currently none requires mechanical ventilation. The child with transglottic stenosis has been decannulated and the one with laryngeal atresia tolerates full capping of his cannula and awaits decannulation. The child with CST had received a tracheostomy for his bronchopulmonary dysplasia. Two years later, the child showed improved lung condition, but the dynamic endoscopy showed lateral collapse of the tracheostoma. This child was decannulated after undergoing a single-stage rib cartilage graft augmentation and external tracheal rigidification with polydiaxanone micro splints.

Discussion

The treatment of TOF is challenging when it is associated with congenital or acquired laryngotracheal stenotic lesions. In this study, 8 of the total 13 patients (61%) with TOF had additional airway anomalies and required concomitant treatment. The laryngotracheal pathologies present in combination with TOF were: laryngeal atresia, laryngeal cleft, suprastomal collapse, subglottic stenosis, transglottic stenosis, long segment cicatricial tracheal stenosis and tracheomalacia. One patient with tracheal agenesis was not offered any treatment, and succumbed to his congenital anomalies.

Diagnosis of TOFs that are recurrent or associated with laryngotracheal lesions is difficult and requires a high index of suspicion [12–14]. They should be suspected in children who had surgery for LTOC (two patients in this series) or OA with TOF (five patients in this series) and continue to have persistent aspiration. Flexible and rigid endoscopy is the gold standard diagnostic modality in such conditions. A tracheoesophageal endoscopy using angled telescopes is critical for defining the fistula demographics that is crucial in selecting the ideal surgical intervention. A correctly performed methylene blue dye test during the endoscopy is a good diagnostic tool to confirm a fistulous tract. The same test is used post-operatively to confirm successful fistula closure. The size of a fistula estimated during endoscopy might be smaller than that observed during the surgery and therefore we emphasize on the importance of performing a meticulous pre-operative endoscopy. Performing the barium swallow test and its interpretation may be difficult in small

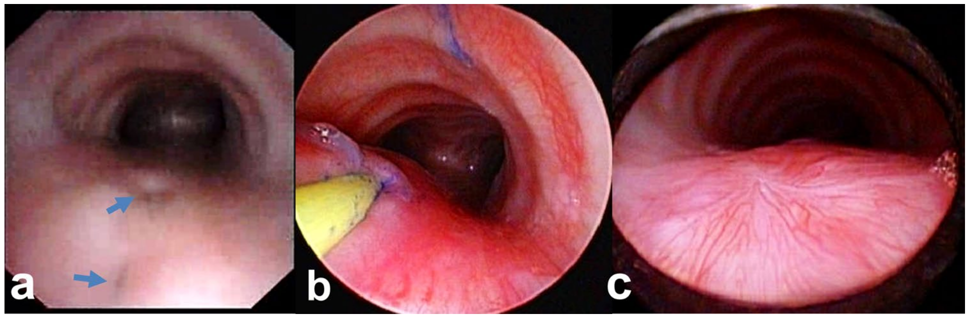

Fig. 2 Endoscopic management of TOF. **a** Flexible endoscopic view of multiple fistulas (blue arrows) in patient # 2. **b** A cytology brush is used to denude the epithelium of the fistula tract. **c** Complete closure of the fistula

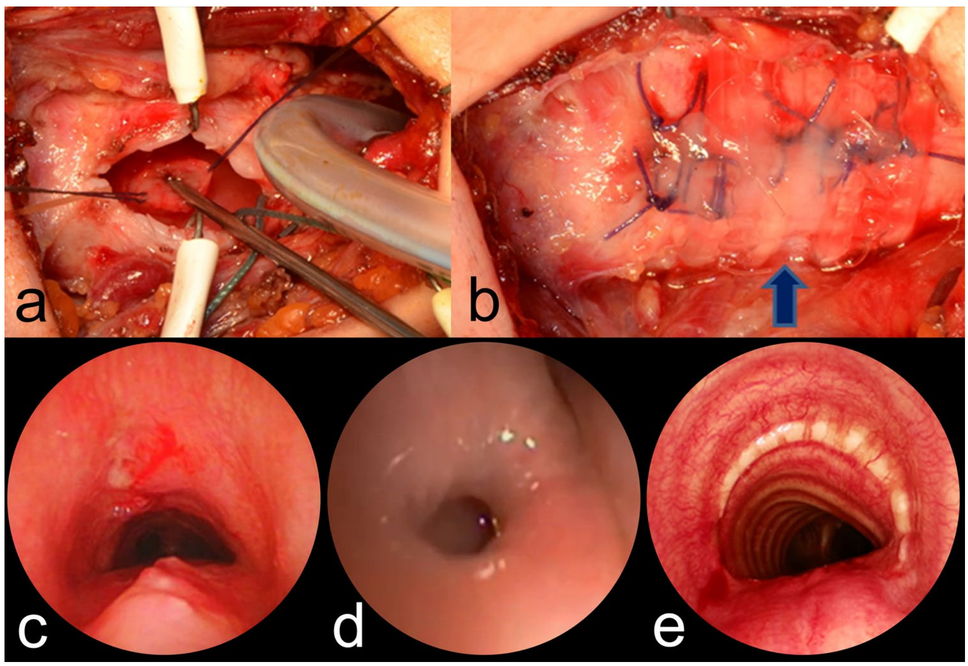

Fig. 3 Surgical repair of TOF. **a** Transtracheal approach to isolate and close the fistula. Note the temporary distal tracheotomy. **b** Closure of the anterior tracheofissure. External resorbable mini splints (blue arrow) rigidified the collapsible tracheal segment. **c** Post-operative view shows successful closure of the TOF. **d** Pre-operative view showing TOF after battery ingestion. **e** Same patient after single stage tracheal resection and anastomosis

children. Therefore, at post-operative follow-ups we rely on parental questionnaires and clinical examination to suspect a recurrent fistula and prefer to confirm it with an endoscopy.

Many techniques (endoscopic and open) have been described for TOF repair to achieve definite healing and prevent recurrences [6, 10]. In our opinion, the success of surgery depends on the fistula etiology and demographics, concomitant aero-digestive pathologies, the choice of surgical technique and the patient's general condition.

Endoscopic techniques are being advocated as an alternative to open repair [7, 8], however, it is necessary to use these techniques in appropriate indications. Endoscopic treatment can be applied on the tracheal and/or oesophageal side of the fistula. One can use electrocautery or lasers, or wire brushes to denude the epithelial tract of the fistula to promote scarring and then occlude it with tissue glue. Endoscopic closure techniques for recurrent TOF are reported to have success rates of 60–84% [15]. We prefer the endoscopic approach for a fistula that is small, easy to expose and has a long slender tract. A large, shallow fistula having a patulous mouth is not amenable for an endoscopic treatment. Surgical dissection and isolation of the fistula trajectory during an OA with TOF repair lengthens the tract. Hence, technically such a fistula is good to treat endoscopically in case of a recurrence. In contrast, a fistula secondary to a failed cleft repair is shallow and requires an open approach. In some cases after denuding the fistulous tract, a transoral endoscopic suturing on the tracheal side of the tract or use of metallic clips to occlude its oesophageal side could be attempted, but this requires special equipment, skills and an optimal collaboration between the surgeon and the anaesthetist. However, following 2 or 3 failed endoscopic attempts, it is best to switch to an open surgery. The use of tracheal or oesophageal stents have been shown to allow fistula closure [16], though in our opinion, it is unsafe to use them in small children. Another advantage of scarring the fistulous tract is the induced cicatricial rigidification of the membranous trachea. This is helpful in patients with a floppy posterior portion of the trachea, as was apparent in one of our patients with Feingold syndrome. Unfavourable cicatrisation of a long LTOC might cause a distal residual TOF and down-sinking of the proximally closed cleft resembling a type I cleft or a deep interarytenoid defect. Endoscopic closure of the proximal residual cleft requires laser incision of the cleft extending above the cuneiform cartilages, followed by a two-layer cleft repair [17]. Distal TOF in such a case is best treated with an open approach for reasons mentioned above.

Open surgery is preferred for a large, residual or recurrent fistula that has failed endoscopic treatment and is associated with a laryngotracheal lesion. TOFs secondary to a caustic injury and battery ingestion are associated with severe inflammation and adhesions and are best managed with open surgery. Concomitant laryngotracheal lesions must be considered while selecting the appropriate surgery for the TOF repair. TOFs associated with severe transglottic stenosis or laryngeal atresia may require an extended cricotracheal resection to simultaneously correct the stenosis and the fistula. In this surgery, the oesophagus is closed vertically and a rib-cartilage graft allows airway expansion and serves as an interpositional tissue between the oesophageal and glotto-tracheal anastomosis. One critical point to remember is to avoid the two anastomotic suture lines from juxtaposing each other to avoid recurrence. A laryngeal drop and adequate cranial mobilisation of the trachea helps in manoeuvring positions of these suture lines. Similar surgical principles must be followed during a tracheal resection and anastomosis.

Patients with OA, isolated TOF and LTOC commonly have moderate to severe degrees of floppy posterior membranous trachea and this can be explained by the developmental theory of these congenital anomalies. A symptomatic child with expiratory dynamic airway collapse could benefit with a posterior tracheopexy performed during the OA and TOF repair, as has been reported by Lawlor et al. [17]. In the treatment of anterior airway malacia, we used external polydioxanone biodegradable micro-splints to rigidify the collapsing tracheal exoskeleton [18]. We used these plates in two patients, relieving one of his respiratory symptoms and allowing decannulation in the other.

Provenzano et al. reported that slide tracheoplasty could successfully repair complex, recurrent and persistent TOFs of different sizes and etiologies [19]. They concluded that the oblique anastomotic line in this technique distributes the tension over a greater surface area and reduces the chances of an anastomotic breakdown. We prefer using this technique in cases of TOF associated with concomitant long tracheal stenosis when an airway resection (more than 5 tracheal rings) and anastomosis would be risky, as was apparent in one of

our patients. In this patient, the oblique tracheal anastomosis avoided juxtaposing over the oesophageal closure. This non-contact between the two suture lines is important to prevent recurrences and in agreement with the Cincinnati groups' report [19]. Some additional techniques have been suggested in open TOF surgeries to prevent recurrences [20, 21]. The sternal or tibial periosteum is quite tough and helps closure of the fistula when interposed between the tracheal and oesophageal walls.

Prevention of TOF formation is important in patients with prolonged endotracheal intubation and tracheostomy. This can be achieved by teaching the pathophysiology of TOF formation to junior doctors, intensive care staff and by ensuring adequate nursing. In cases of LTOC and LA, the site of performing a tracheostomy (if deemed necessary) is crucial. In a LTOC, the tracheostomy should be performed at least two rings distal to the cleft. This will avoid constant friction between the cannula and the cleft repair and thus prevent formation of a residual fistula at its lower end. It will also minimise the risk of infection at the cleft repair site and reduce the risk of breakdown. Patients with LA usually have a proximal fistula. These patients present with congenital high airway obstructive syndrome (CHAOS), are delivered with the EXIT procedure and undergo tracheostomy before separating the feto-maternal circulation. Tracheostomy in these cases should be performed either close to the cricoid or much distally between the 6th and 7th tracheal rings. In future, this will then allow an appropriate combined management of the agenesis and the fistula without needing to excise additional tracheal rings during the extended cricotracheal resection.

We observed that associated laryngeal and tracheal anomalies, the fistula etiology and its size influenced the choice of the surgical management and had an impact on the treatment outcome. These are rare cases and the existing literature on their management is diverse. A single institution does not experience many such patients and therefore, ours was a small case series, and the patient groups were heterogenous. Until large multi-centric comparative studies are available, the choice of surgical treatment might be biased and influenced by the training received by the surgeon. Further studies are required to establish a systematic protocol for managing these challenging cases and validate their results.

Conclusion

A pre-operative endoscopic evaluation is important for selection of the appropriate surgery for TOF to prevent its recurrence and operative complications. One must know about the indications and limitations of the endoscopic approaches for TOF repair. Several open approaches are available and their selection is based on the etiology and demographics of the fistula and the patient's general condition. Concomitant management of associated laryngotracheal lesions is critical.

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Compliance with ethical standards

Conflict of interest None of the authors have any conflicts of interest.

Human and animal rights Yes.

Informed consent Yes.

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