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Esophageal atresia repair with thoracotomy: the Cincinnati contemporary experience

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Esophageal atresia/tracheoesophageal fistula (EA/TEF) repair using an open muscle-sparing thoracotomy has been the standard approach used in our institution. Whereas perioperative mortality is now very uncommon, short- and long-term morbidity is very common in these patients. However, the complexity of the esophageal anatomy and significant comorbidities appear to be important contributors to significant complications in these patients. At least 30% of the EA/TEF patients required esophageal dilatations for anastomotic stricture; this increased to 50% for patients with pure EA. Gastroesophageal reflux requiring an antireflux procedure was performed 23% of the time for EA/TEF and 30% for EA patients. In addition, there were a few complications, such as winging of the scapula and scoliosis, that were attributed in part to the utilization of a nonmuscle-sparing thoracotomy. The standard muscle-sparing thoracotomy remains a very versatile and useful approach to repairing esophageal atresia, and it is the standard for repairing more complex anatomical variants. The self-reported long-term quality of life in these patients is very good, except for a few individuals with protracted feeding disorders and severe dysphagia.

Esophageal atresia and tracheoesophageal fistula (EA/TEF) is a relatively rare condition, affecting approximately 1 in 2500 to 4500 live births.[1,2] Although EA is an infrequent occurrence, its complex nature still poses significant challenges for the pediatric surgeon charged with the care of these children today.[3] The recent improved survival of these neonates is dependent on a number of factors, including advances in neonatal intensive care and anesthesia as well as improved surgical techniques, parenteral nutrition, and antibiotics. Even more challenging than the actual initial corrective surgery is the management of the complications, which can ensue in the early or late postoperative phase. These require a repertoire of surgical skills and treatment strategies to reduce the risk of long-term adverse sequellae.

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In this paper, we will endeavor to outline the potential complications following EA repair in a large North American children's hospital, and we will also discuss the various methods and approaches that may be used to manage the complications and sequella of EA/TEF repair.

Materials and methods

We performed a retrospective review of the charts of all patients treated operatively in Cincinnati Children's Hospital Medical Center (CCHMC) with a diagnosis of EA over a 10-year period, from 1997 to 2007 inclusive. All patients underwent an initial assessment of their physiological status preoperatively, including the degree of prematurity and respiratory and cardiovascular status before any surgical intervention. An initial screen for major anomalies included a full physical examination and a chest radiograph to confirm the diagnosis and assess the heart and lungs. Further studies

included an echocardiogram (to establish the presence or absence of structural anomalies and the location of the aortic arch) and a renal ultrasound, to determine whether renal anomalies were present or not.

Initial management

Physiologically unstable or very premature infants (<1500 g) underwent initial stabilizing procedures, such as central line insertion and, in some cases, a TEF ligation and open (Stamm) gastrostomy tube insertion. These patients subsequently had delayed primary repair of their EA when stable and once their weight was >2000 g. In infants who were initially stable, a bronchoscopy was performed in the operating room under general anesthesia, and a primary repair was performed within 48 hours after birth. Patients who were diagnosed with a pure EA without a fistula all underwent delayed repair, after a prolonged period of proximal esophageal pouch Replogle suctioning and upper pouch bouginage in an effort to allow esophageal lengthening and growth.

Surgical approach

A number of parameters were looked at during the initial bronchoscopy to guide further surgical management. The important features noted were the fistula location and whether one, two, or more fistulae[4] could be identified. The presence or absence of a laryngo-esophageal cleft was also looked for, as well as any evidence of vascular compression of the trachea. The default operative approach in most cases was via a muscle-sparing right thoracotomy. If a right-sided aortic arch was found on preoperative echocardiography, then a left thoracotomy was employed.[5] In most cases, an extra-pleural approach was utilized to gain exposure to the EA. The TEF was ligated initially, followed by mobilization of the proximal pouch and subsequent mobilization of the distal esophageal segment. The esophageal ends were then anastomosed in a single layer with a monofilament absorbable suture to achieve esophageal continuity. A small number of patients with lethal chromosomal, neurological, or a combination of nonsurvivable anomalies were deemed unfit for surgical intervention and were not included in our study.

Results

A total of 92 patients, with a diagnosis of EA, were treated by the pediatric surgical faculty at CCHMC between the years of 1997 and 2007, and their accessible records were reviewed. The dates of birth for these patients ranged from 1985 to 2007. Of these 92 patients, 78 were treated from birth at this institution, and 14 patients were referred well after the neonatal period for treatment from other centers. All patients had a thoracotomy for definitive repair of their EA.

The spectrum of EA and TEF anomalies treated in our institution broadly reflect the known distribution of the various subtypes, with EA and distal TEF comprising 70 patients (76.1%); pure EA affecting 16 patients (17.3%); EA with a proximal TEF occurring in 3 patients (3.3%); and 3 patients having EA with both proximal and distal TEFs (3.3%). A long-gap atresia (>2.5 to 6 vertebral body gap) was noted in 26 patients (28%) in our study.

Of the 70 patients with EA and a distal TEF operated on at CCHMC, a significant number had associated anomalies (Table 1). Seven of the patients (10%) were born prematurely. Congenital heart disease was present in 21 patients (30%); genito-urinary (GU) and central nervous system (CNS) anomalies were present in 7 patients each (10%); duodenal atresia and vertebral anomalies were present in 6 patients each (8.5%); imperforate anus and chromosomal anomalies were present in 4 patients each (6%); and laryngo-tracheal cleft and limb anomalies were present in 3 patients each (4%).

Of the 16 patients with pure EA, 3 of the patients (18%) were born prematurely. The other associated anomalies found in this group included congenital heart disease in 5 patients (31%), chromosomal and CNS anomalies in 3 cases each (19%), with Trisomy 21 being the chromosomal abnormality in all 3 cases. The other anomalies included imperforate anus and duodenal atresia in 2 cases each (12.5%) and GU anomalies in 1 patient (6%).

Table 1 Anomalies associated with esophageal atresia

	EA/distal TEF (n = 70)	Pure EA (n = 16)	EA/proximal TEF (n = 3)	EA/proximal & distal TEF (n = 3)
Congenital heart disease	21 (30%)	5 (31%)	1 (33%)	1 (33%)
CNS anomalies	7 (10%)	3 (19%)	—	—
Prematurity	7 (10%)	3 (18%)	1 (33%)	1 (33%)
GU anomalies	7 (10%)	1 (6%)	—	—
Vertebral anomalies	6 (8.5%)	—	—	—
Duodenal atresia	6 (8.5%)	2 (12.5%)	—	—
Imperforate anus	4 (6%)	2 (12.5%)	—	—
Chromosomal anomalies	4 (6%)	3 (19%)	—	—
Laryngotracheal cleft	3 (4%)	—	—	—
Limb anomalies	3 (4%)	—	—	—

Of the three patients with an EA and a proximal TEF, one patient was born prematurely (33%) and one had an associated congenital cardiac anomaly (33%). In two of these three patients, the proximal TEF was missed initially. There were similar findings in the group of three patients with EA and both a proximal and distal TEF, with one patient (33%) being born prematurely and one patient (33%) having cardiac anomalies. In this group, the proximal TEF was missed initially in one patient.

A definitive surgical repair was possible in the majority of the 70 patients with EA and a distal TEF. A staged repair was required in a small number of cases, and these involved a proximal pouch myotomy in 4 cases (5.8%), colonic interposition in 7 cases (10%), a reverse gastric tube in 2 cases (2.9%), and a gastric transposition in 1 patient (1.4%).

The complication rates were comparable to the known complications following EA repair.[6] There were no perioperative deaths in this group. An anastomotic leak can be a devastating complication and may result in mortality as a result of mediastinitis and irreversible sepsis. Many surgeons routinely leave a chest tube (extrapleural or intrapleural) in position, until a leak has been ruled out by a contrast esophagram, to prevent the sequellae of this problem. Three patients in our series developed a minor leak (one occurred after a dilation), and two patients had a major leak or partial esophageal dehiscence (Table 2). Surgical intervention was not required for the minor leaks as these healed with ongoing conservative management. However, 22 patients (31%) developed an esophageal stricture, which required dilation. An average of 4 dilatations was required per patient, with a range of 1 to 10 dilatations being required overall. Despite 31% of patients having a stricture requiring dilation, only 2 (2.9%) needed actual resection of the stricture.

Recurrence of a TEF can be a difficult problem to diagnose and manage, and this complication occurred in five patients (7%). All of these patients underwent reoperation with fistula obliteration. Three of these patients underwent an endoscopic repair of the recurrent fistula, and two patients had an open repair.

Management of the neonatal airway poses many challenges, and in seven children (10%), a tracheostomy was required for reasons such as subglottic stenosis, bilateral vocal cord paralysis, tracheomalacia, or laryngotracheoesophageal cleft (LTC). Four patients (5.8%) developed subglottic stenosis in the postoperative period, and although this complication may not be directly related to the operative repair of the EA/TEF, it most commonly results from prolonged postoperative intubation and ventilation and is a significant morbidity. Two patients (2.9%) developed a bilateral vocal cord paralysis.

Table 2 Complications associated with esophageal atresia repair

	EA & distal TEF (n = 70)	Pure EA (n = 16)
Stricture	22 (31%)	8 (50%)
Recurrent TEF	5 (7%)	—
Subglottic stenosis	4 (5.8%)	—
Minor leak	3 (4.3%)	2 (12.5%)
Major leak (dehiscence)	2 (2.9%)	1 (6.7%)
Stricture resection	2 (2.9%)	2 (12.5%)*
Perioperative death	—	—

*Foker procedure.

Table 3 Additional procedures post-repair of esophageal atresia

	EA & distal TEF (n = 70)	Pure EA (n = 16)
Esophageal dilatations	22 (31%)	8 (50%)
CHD, PDA, vascular ring repair	21 (30%)	5 (31%)
Fundoplication	16 (23%)	5 (31%)
Esophageal foreign body removal	8 (11.4%)	—
Tracheostomy	7 (10%)	3 (18.75%)
Duodenoduodenostomy	6 (8.5%)	2 (12.5%)
Aortopexy	4 (5.7%)	—
Imperforate anus repair	4 (5.7%)	2 (12.5%)

Due to the complexity of these patients, a number of other procedures were often required (Table 3). These may be due to complications directly related to the EA and TEF or were due to other significant medical problems and comorbidities that come to light during the initial preoperative workup or subsequently during their prolonged hospital stay. A large number of these children had symptomatic gastro-esophageal reflux with or without an esophageal stricture, and therefore 16 patients (23%) required a fundoplication. Because of the regular occurrence of esophageal strictures postoperatively, a number of children re-present with bolus obstruction of the esophagus. Eight children (11.4%) had coins removed endoscopically from their esophagus. An aortopexy was required in 4 children (5.7%) for the management of life-threatening tracheomalacia. Cardiovascular anomalies that required surgical correction included congenital heart disease, patent ductus arteriosus (PDA), and vascular rings in 22 patients (31%). Duodenal atresia and imperforate anus was present in 6 patients (8.5%) and 4 patients (5.7%), respectively.

Delayed primary repair was possible in nine cases (56%) of pure EA. The difficulty in achieving a tension-free anastomosis is apparent from the number of alternative techniques employed to achieve esophageal continuity. A colonic interposition was utilized in five patients (31%); a proximal esophageal myotomy was performed in two patients (12.5%); and a reverse gastric tube was created in one case (6%).

The complications occurring in patients with pure EA were significantly higher, primarily due to the difficulty in getting continuity without tension. There were no perioperative deaths in the pure EA group. Eight of these patients (50%) developed a stricture, and all of those developing a stricture required esophageal dilatations. An average of 4 dilatations was required per patient with a range of 1 to 12

being required overall. Two patients (12.5%) required operative intervention for their stricture with an anastomotic resection using a Foker technique.[7,8] Two patients (12.5%) developed a minor leak, and one patient (6.3%) had a major esophageal leak.

A number of additional procedures were required for this group of patients, with a fundoplication being the most commonly employed operation in five patients (31%), due to the high incidence of gastro-esophageal reflux and its detrimental effects on esophageal strictures. Five patients (31%) underwent cardiac surgery, involving repair of a PDA, vascular ring, or other congenital heart condition. Two patients (12.5%) underwent repair of duodenal atresia, and two patients (12.5%) underwent repair of an imperforate anus. A tracheostomy was performed in three patients (18.8%).

In total, 26 patients were noted to have a long-gap EA with or without TEF, where the esophageal gap was at least 2.5 and 6 vertebral bodies. A delayed primary anastomosis was possible in 14 of these patients (54%). To achieve a primary anastomosis, a myotomy was performed in 6 cases (23.1%). Of these 6 cases, 3 developed a leak and 3 subsequently required a stricture resection. The Foker procedure for esophageal lengthening was utilized in 1 case, and a subsequent colon interposition was necessary in 2 cases.

Colonic interposition was widely used as the first procedure in the long-gap group, and it was performed in 10 cases (38%). However, a reverse gastric tube was only used in two cases (8%) as a first procedure with one of these requiring a colon interposition as a second procedure.

Our patients have undergone long-term follow-up from 1 to 27 years with a median follow-up of 6 years, with a view to assessing their long-term outcomes. Three patients (3%) died at 1, 3 and 12 years, respectively, all of which were unrelated to their EA.

Dysphagia after age 5 was a common problem, affected at least 11 patients of 46 (24%), who were followed long term. The degree of dysphagia was felt to be severe in 3 cases, with 1 patient still having oral aversion and feeding disorder at age 7 years. In 8 cases, the dysphagia was classified as mild and intermittent and did not appear to affect the overall nutritional status and quality of life of the patients.

Congenital and acquired chest wall and spinal anomalies have been associated with EA and TEF. These defects have been linked to underlying vertebral anomalies as part of the VACTERL association or may also be due to injury to scapula musculature during thoracotomy and distortion of ribs after healing. At long-term follow-up, 9 patients out of 46 (20%) were noted to have some chest wall or spinal deformities. Two patients had developed pectus excavatum, and 3 patients were noted to have significant scoliosis. Chest wall asymmetry or winging of the scapula occurred in 4 cases, which was commonly attributed to the use of a nonmuscle-sparing incision used at thoracotomy.

The quality of life for most patients was self-rated as excellent and very good. The exceptions were a few patients with protracted feeding problems and severe dysphagia.

Discussion

Surgery for children with EA and TEF has always been challenging and has several known potential complications. These postoperative problems can occur despite the favorable anatomy that may be encountered in a short-gap EA and despite meticulous surgical technique with excellent postoperative management. The demanding nature of this surgery is such that morbidities unfortunately occur and must be dealt with. More importantly, they should be recognized as early as possible and definitive therapy instituted in order that poor long-term outcomes can be avoided. By discussing our experience of patients with EA and TEF and the potential pitfalls, we hope to clearly delineate the warning signs that should be picked up.

In general, complications can arise as an early or a late occurrence. The early complications are associated with both surgical techniques as well as certain patient factors that in turn may compound the effect of surgical technique. These complications include: anastomotic leaks (radiological or incidental, minor leak, and major leak), anastomotic stricture, recurrent TEF, and esophageal dysmotility with an associated risk of aspiration.

Incidental or radiological leaks are those that are identified on a routine postoperative contrast study before the commencement of oral feeds. This finding is usually not of any significance and can be treated expectantly, with the vast majority healing within a few days.[9] The mainstay of treatment is the avoidance of oral feeds with the institution of either parenteral nutrition or trans-anastomotic tube (TAT) feeds. If a chest tube (extrapleural or intrapleural) had been placed at the time of operation, it is maintained until the leak is closed. Since extrapleural chest tubes are not routinely placed by every surgeon, a very small leak of saliva from the anastomosis does not always need to be drained and any evidence of a significant pneumothorax would require a separate intrapleural thoracostomy tube.[1] When leaks occur, saliva may or may not come from the chest tube, depending on the extent of the leak. Antibiotics and suctioning of the upper pouch may be instituted to reduce saliva egress from the esophagus. A repeat contrast study before oral feeding is at the discretion of the surgeon.

A minor leak, which occurs in at least 6% to 17% of cases,[10,11] usually presents with saliva in the chest tube also and can be managed in a similar manner to that mentioned previously. Factors that may contribute to leaks include poor suture technique, with too few or even too many sutures being placed. Knots tied too tightly or with the mucosa excluded from the anastomosis have also been proposed to lead to poor anastomotic healing. Some animal studies have suggested that knots left on the luminal aspect

of the esophagus increase the risk of subsequent stricture.[12] Tension on the suture line is obviously a contributing factor to the possibility of a leak and subsequent stricture formation. In order that tension can be reduced, it is possible to mobilize both proximal and distal esophageal pouches,[13] and this is often tempting. However, excessive mobilization of the esophagus with inadvertent devascularization and subsequent ischemia of the esophagus can clearly impair the ability of the anastomosis to heal without subsequent fibrosis, scarring, and stricture formation. The postoperative intensive care of these infants is at least as important as the surgical correction of their anatomical abnormality. If vigorous neck extension is employed during attempts at reintubation or if injudicious passage of a nasogastric tube is performed, then the anastomosis may be partially or completely disrupted in the process.

A major leak (3-5%) is a potentially disastrous consequence and will usually occur early in the postoperative course (within 48-72 hours). These infants may develop symptoms and signs of mediastinitis. An acute life-threatening episode may signify the development of a tension pneumothorax and an inability to ventilate and oxygenate, which requires immediate needle decompression, followed by tube thoracostomy. An empyema or mediastinal abscess may form if the leak goes unnoticed for a prolonged period of time, and this requires drainage of the collection with administration of systemic broad-spectrum antibiotics until complete resolution has occurred. Attempts to re-anastomose the disrupted esophagus may be initially unsuccessful as the friability of the tissues can preclude a satisfactory re-anastomosis. Some surgeons have advocated early re-exploration, as the placement of a few additional sutures may be all that is required.[1] Alternatively, a safe course of action is creating a cervical esophagostomy and a gastrostomy to simply drain and defunctionalize the esophagus. This procedure can seem counter-productive initially, but may in fact be life-saving in the face of a very ill and septic neonate. Unfortunately, the incidence of esophageal stricture following an anastomotic leak is significantly higher.

The incidence of anastomotic stricture varies widely and is the most common cause of recurrent surgery in children with EA and TEF. The majority of studies report a stricture rate of between 37% and 52%[1,14]; however, one study quoted rates as high as 69%.[6] The definition of a stricture, however, is not universally accepted or agreed, as a mild radiological narrowing on a contrast esophagram may not have any clinical relevance to the physician or the patient (who may be able to swallow satisfactorily), and therefore the rates may not always be comparable. A number of factors can predispose toward stricture formation and should be avoided, if at all possible. Significant tension on the anastomosis, with resultant low-grade ischemia of the proximal and distal ends of the esophagus, is commonly felt to be the most obvious cause of a stricture. Obviously, careful handling of the thin, delicate tissue during mobilization of the neonatal esophagus is a basic surgical requirement. A contributing factor to possible stricture formation can be the suturing technique that is employed. A two-layered anastomosis may often be reassuring as it can redistribute some of the direct forces on the friable esophageal tissues on either side of the anastomosis; however, it also increases the amount of tension applied to the remaining esophagus and may be detrimental to intramural vascularity in the long run. The two-layered anastomosis is associated with less initial leaks but a slightly higher rate of strictures.[15] For this reason, many surgeons today favor a single-layer anastomosis with fewer rather than more sutures, and in general, 6 to 8 interrupted sutures may be all that is required. It has been postulated that silk sutures, although very easy to handle and tie, may incite an ongoing inflammatory reaction over a long period of time and may actually increase the incidence of stricture compared with long-lasting monofilament absorbable sutures.[12]

An interesting paper by Carachi and coworkers[12] looked at the effect of an indwelling silicone trans-anastomotic tube (TAT) on the healing esophageal anastomosis in canine pups. They found no significant difference in stenosis rates whether a TAT was used or not. Interestingly, they observed a shelf of stenotic tissue on the posterior wall of the esophagus at the site where the intra luminal silk knots had been tied, compared with a thin linear scar on the anterior wall, where knots had been tied extraluminally.

Gastroesophageal reflux (GER) is extremely common among babies following EA repair and may affect between 40% and 65% of patients.[16,17] The presence of significant GER is generally believed to be due in part to an intrinsic deficiency in the motor function of the esophagus itself.[18] However, it is likely that GER is exacerbated by the surgical repair and gastrostomy, causing an alteration of the anatomical gastro-esophageal junction and the angle of His. Although, the majority of these patients can be treated medically with H_2-receptor blockers or proton-pump inhibitors,[14,19] about 28% of them will require surgical correction of their reflux.[20] The diagnosis of reflux is confirmed with a contrast swallow, esophagoscopy with biopsy, or 24-hour pH probe. An associated stricture can also be determined at this time, either radiographically or by visual inspection for signs of esophagitis. A biopsy of the inflamed esophagus can yield a histopathological diagnosis of reflux esophagitis in 20% of cases and Barrett's esophagus in 6%.[21] Dilation of the stricture can be performed by various forms of bouginage (Maloney, filiform, or Tucker) or preferably with balloon dilation under fluoroscopic or endoscopic control. Most strictures respond to dilation, but it is crucial that reflux is aggressively treated to diminish the impact of acid reflux for recurrent stricture formation. In our experience, almost 30% of the patients required a fundoplication. A short, loose Nissen fundoplication is the procedure of choice, but unfortunately, a significant number of infants (>40%) will develop recurrent GER[22] and this may in part be due to the inherent dysmotility of the esophagus.[23] In children who do not respond to esophageal dilatations, ongoing

GER is the commonest cause for their failure to improve. As a last resort, resection of the affected esophageal segment may be required, but in general this is uncommon.

A recurrent TEF occurs in 3% to 15% of cases, and it usually is located in the region of the original fistula.[6,15] The incidence of recurrent TEF is much higher following an anastomotic leak, and this may often have resulted from excessive tension during the anastomosis.[24] The majority of recurrent TEFs present early but some can occur months and even years after the initial repair. The presenting symptoms usually involve coughing, choking, cyanosis with feeding, and/or recurrent chest infections or pneumonia.[25] The diagnosis is made either by direct visualization of the fistula during bronchoscopy or during a contrast swallow, when contrast may be seen entering the airway from the esophagus, although both of these studies can miss a recurrent fistula.[26] In general, spontaneous closure of the fistula is unusual and surgery is required. This should preferably be deferred for a period of time (up to 1 month) if the child's clinical condition allows, as early re-operation is particularly challenging. During this period, nutrition can be optimized. The standard surgical approach involves a thoracotomy, and this surgery is notably difficult with potential associated morbidity,[27] including a 10% to 22% incidence rate of recurrent fistula.[15,26] Identification of the fistula is made easier with the passage of a catheter through the fistula at bronchoscopy.[28] In view of the risks of open surgery, certain authors have advocated the use of minimally invasive techniques (either bronchoscopic or esophagoscopic) to deal with the recurrent TEF.[29,30] However, many of the tissue adhesives used (Histoacryl, fibrin glue) have been of limited success and, despite multiple applications, have resulted in poor fistula closure rates.[31,32] Rutter and colleagues at our institution have successfully used an endoscopic cautery device to coagulate the recurrent fistula track followed by placement of a tissue adhesive in several patients with recurrent TEF.[33] With few exceptions, this is our preferred method of initial management today.

Long-term outcome issues and complications arise as a result of other anomalies and comorbidities associated with EA and therefore may be difficult to manage. These anomalies are most significant in neonates born with the VACTERL association. The VACTERL association is a spectrum of clinical conditions in the human neonate, which involves multiple anomalies first reported by Quan.[34] The exact incidence of VACTERL is difficult to quantify in view of the fact that there is such variation in clinical presentation, but it affects approximately 1 in 5000 live births. VACTERL has not been recognized as a specific syndrome in humans but rather represents a nonrandom association of congenital anomalies of poorly known etiology and pathogenesis and its components have been variable. The most important features of the VACTERL association include vertebral anomalies (V), anorectal malformations (A), cardiac anomalies (C), tracheo-esophageal fistula (T), esophageal atresia (E), renal anomalies (R), and limb problems (L). Etiologically, the VACTERL association may be a feature of some chromosomal anomalies,[35] but the majority of cases have no recognized cause. If major cardiac, neurological, or urological anomalies are present at birth, they may adversely affect the patient's outcome in a number of ways. This is not only of relevance in the initial postoperative recovery phase but also plays a role in some infants' suitability for the initial surgery. Our data came from all neonates who underwent initial workup and were found not to have a lethal anomaly that would preclude them from surgery.

Tracheomalacia is a common finding among neonates with EA and often manifests itself as the classic "TEF cough," which can persist into adult life. Significant tracheomalacia, however, is present in only 10% to 20% of infants, with even fewer requiring surgical intervention.[36] In general, tracheomalacia improves with age,[37] regardless of whether it is treated or not. Bronchoscopy is the gold standard for diagnosis, with bulging of the posterior tracheal wall being the cardinal feature, except in severe cases where near-total anteroposterior collapse is evident.[38] Surgery is reserved for those with near-death episodes or recurrent pneumonia and involves an aortopexy to lift the anterior wall of the trachea forwards. Success rates in the region of 35% to 88% have been achieved with this technique.[39,40]

Esophageal dysmotility is a very common long-term finding in children with EA/TEF and has been demonstrated in 75% to 100% of patients post-EA/TEF repair.[21] Likewise, in patients who have had some form of esophageal replacement, dysmotility with symptoms such as aspiration, dysphagia, or food bolus obstruction is often experienced.[41] In animal studies, dogs have been widely used to study the postoperative results following esophageal resection or transection with subsequent re-anastomosis, utilizing an autologous jejunal mucosa transplant,[42] a tubular musculopleural pedicle graft,[43] or a variety of myotomies with or without delayed esophageal reconstruction.[44,45] The results from these studies were variable with no clear advantage for one particular technique. Although early contrast and manometric studies suggested a good outcome from spiral myotomy, long-term follow-up in this group demonstrated esophageal dysmotility, as has also been observed in children following esophageal surgery. However, further studies in canine models of EA compared manometric findings following esophageal transection and re-anastomosis versus esophageal vagotomy alone.[46] Their results showed coordinated peristaltic contractions between the proximal and distal esophagus in the first group with abnormal simultaneous contractions in the vagotomy group. This suggested that postoperative dysmotility might arise from disruption of the vagus nerve either as a part of the congenital abnormality or secondary to surgical trauma.

The topic of esophageal replacement has always prompted vigorous debate as to what is the best conduit for children with little or no esophagus. Obviously the best esophageal conduit is the child's native esophagus and no other substitute is truly ideal. The most common alternative

conduits include stomach (gastric transposition or gastric tube), colon, and jejunum. The stomach is useful for a number of reasons, including only a single anastomosis in the neck or chest and having a good blood supply with decreased risk of ischemia, leak, and stricture. Conversely, it will occupy a large space in the chest and may promote reflux with the associated risks of delayed emptying due to a vagotomy. A large series by Spitz performed 175 gastric pull-ups over 21 years with no graft failures but 9 deaths (5.2%).[47] Anastomotic leaks occurred in 12% and 19% of patients developed strictures requiring dilation. A good functional outcome was achieved overall.[48]

A gastric tube can be constructed as either a reversed or isoperistaltic tube from the greater curvature of the stomach. The blood supply is generally excellent, and it tends to retain its tubular shape. It does, however, reduce the stomach capacity, and the tube will produce acid long term. Complications such as anastomotic leak in 50% of cases and stricture formation in 66% of cases seem significant, but the long-term outcome was reasonable in terms of swallowing.[49]

Colonic transposition is the most frequently used substitute for the esophagus. It is relatively easy to perform the surgery, but there are multiple anastomoses with an increased risk of a leak and the upper anastomosis is prone to ischemic strictures. Over time the colon can become tortuous, resulting in stasis. A very large series from Egypt reported 775 colon interpositions over a 30-year period (mostly for caustic strictures) with excellent results. Only 10% of the upper anastomoses leaked with a 5% proximal stricture rate and an overall mortality of 1%.[50] Chronic gastrocolic reflux can lead to ulceration. In general, it has been shown to be safe and yields satisfactory results over time.[51]

Pedicled or free jejunal or ileal graft interposition has been employed with some success[52]; however, significant complications occur on a frequent basis. Graft necrosis, ischemia, strictures, and death are the most significant complications and occur not infrequently.[53,54]

Chest wall and spinal deformities can be very disfiguring for patients and may be inadvertently overlooked from a pediatric surgeon's point of view, as they may only become apparent in later life and may be referred to a different specialty, such as orthopedics or plastic surgery. Open thoracotomy can result in significant musculoskeletal morbidity if care is not taken to ensure proper muscle-sparing surgical technique. Associated vertebral anomalies can contribute to the chest wall or spinal deformity by a direct effect on the ribs and vertebral column. A "winged" scapula secondary to neuromuscular injury to the latissimus dorsi muscle has been reported in 24% of patients[55] undergoing a standard posterolateral thoracotomy for EA/TEF repair with up to 21% of patients having a scoliosis.[21] Scoliosis was more common in patients who had undergone more than one thoracotomy or division of portions of the serratus anterior and latissimus dorsi muscle groups or their nerve supply. Some females were found to have developed breast asymmetry and elected to have reconstructive surgery.[56] In response to some of these problems, Bianchi developed a high axillary skin crease incision in an attempt to improve esthetics and to reduce the incidence of musculoskeletal deformity.[57,58]

With the open thoracotomy approach used in our institution, perioperative mortality is now very uncommon. Although short- and long-term morbidity is very common in these patients, few of the complications seen are related to the current surgical approach. The complexity of anatomy and significant comorbidities are generally much more important. Proximal TEF and laryngotracheoesophageal clefts (grade 1 and 2) may be missed without prerepair endoscopy, and therefore it is vital that they are carefully looked for.

In our experience, circular myotomy is associated with a 50% leak rate and eventual failure requiring esophageal replacement in 33% of cases. Dysmotility and dysphagia occur in almost all of these patients, and therefore, we no longer recommend the technique of myotomy for patients with EA.

Quality of life is very important for the long-term well being of patients who have undergone EA/TEF repair, and certain studies have shown that quality of life in adults after EA/TEF repair is comparable with that of healthy adults in the majority of cases.[59,60] The standard muscle-sparing thoracotomy remains a very versatile and useful approach to repairing EA, and it is the standard for repairing more complex anatomical variants.

References

1. Spitz L. Esophageal atresia. Lessons I have learned in a 40-year experience. J Pediatr Surg 2006;41(10):1635-40.
2. David TJ, O'Callaghan SE. Oesophageal atresia in the South West of England. J Med Genet 1975;12(1):1-11.
3. Myers NA. Oesophageal atresia: the epitome of modern surgery. Ann R Coll Surg Engl 1974;54(6):277-87.
4. 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.
5. Allen SR, Ignacio R, Falcone RA, et al. The effect of a right-sided aortic arch on outcome in children with esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 2006;41(3):479-83.
6. Kovesi T, Rubin S. Long-term complications of congenital esophageal atresia and/or tracheoesophageal fistula. Chest 2004;126(3):915-25.
7. Till H, Muensterer OJ, Rolle U, et al. Staged esophageal lengthening with internal and subsequent external traction sutures leads to primary repair of an ultralong gap esophageal atresia with upper pouch tracheoesophagel fistula. J Pediatr Surg 2008;43(6):E33-5.
8. Foker JE, Kendall TC, Catton K, et al. A flexible approach to achieve a true primary repair for all infants with esophageal atresia. Semin Pediatr Surg 2005;14(1):8-15.
9. Chittmittrapap S, Spitz L, Kiely EM, et al. Anastomotic leakage following surgery for esophageal atresia. J Pediatr Surg 1992;27(1):29-32.
10. Lilja HE, Wester T. Outcome in neonates with esophageal atresia treated over the last 20 years. Pediatr Surg Int 2008;24(5):531-6.
11. Orford J, Cass DT, Glasson MJ. Advances in the treatment of oesophageal atresia over three decades: the 1970s and the 1990s. Pediatr Surg Int 2004;20(6):402-7.
12. Carachi R, Stokes KB, Brown TC, et al. Esophageal anastomosis–an experimental model to study the anastomotic lumen and the influence of a transanastomotic tube. J Pediatr Surg 1984;19(1):90-3.

13. Lessin MS, Wesselhoeft CW, Luks FI, et al. Primary repair of long-gap esophageal atresia by mobilization of the distal esophagus. Eur J Pediatr Surg 1999;9(6):369-72.
14. Konkin DE, O'Hali WA, Webber EM, et al. Outcomes in esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 2003; 38(12):1726-9.
15. Engum SA, Grosfeld JL, West KL, et al. Analysis of morbidity and mortality in 227 cases of esophageal atresia and/or tracheoesophageal fistula over two decades. Arch Surg 1995;130(5):502-8, discussion 508-9.
16. Curci MR, Dibbins AW. Problems associated with a Nissen fundoplication following tracheoesophageal fistula and esophageal atresia repair. Arch Surg 1988;123(5):618-20.
17. Jolley SG, Johnson DG, Roberts CC, et al. Patterns of gastroesophageal reflux in children following repair of esophageal atresia and distal tracheoesophageal fistula. J Pediatr Surg 1980;15(6):857-62.
18. Romeo G, Zuccarrello B, Proiette F, et al. Disorders of the esophageal motor activity in atresia of the esophagus. J Pediatr Surg 1987;22(2): 120-4.
19. Van Biervliet S, Van Winckel M, Robberecht E, et al. High-dose omeprazole in esophagitis with stenosis after surgical treatment of esophageal atresia. J Pediatr Surg 2001;36(9):1416-8.
20. Little DC, Rescorla FJ, Grosfeld JL, et al. Long-term analysis of children with esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 2003;38(6):852-6.
21. Somppi E, Tammela O, Ruuska T, et al. Outcome of patients operated on for esophageal atresia: 30 years' experience. J Pediatr Surg 1998; 33(9):1341-6.
22. Kubiak R, Spitz L, Kiely EM, et al. Effectiveness of fundoplication in early infancy. J Pediatr Surg 1999;34(2):295-9.
23. Boleken M, Demerbilek S, Kirimiloglu H, et al. Reduced neuronal innervation in the distal end of the proximal esophageal atretic segment in cases of esophageal atresia with distal tracheoesophageal fistula. World J Surg 2007;31(7):1512-7.
24. Vos A, Ekkelkamp S. Congenital tracheoesophageal fistula: preventing recurrence. J Pediatr Surg 1996;31(7):936-8.
25. Ghandour KE, Spitz L, Brereton RJ, et al. Recurrent tracheo-oesophageal fistula: experience with 24 patients. J Paediatr Child Health 1990;26(2):89-91.
26. Ein SH, Stringer DA, Stephens CA, et al. Recurrent tracheoesophageal fistulas seventeen-year review. J Pediatr Surg 1983;18(4):436-41.
27. Myers NA, Beasley SW, Auldist AW. Secondary esophageal surgery following repair of esophageal atresia with distal tracheoesophageal fistula. J Pediatr Surg 1990;25(7):773-7.
28. Atzori P, Iacobelli BD, Botterro S, et al. Preoperative tracheobronchoscopy in newborns with esophageal atresia: does it matter? J Pediatr Surg 2006;41(6):1054-7.
29. Hoelzer DJ, Luft JD. Successful long-term endoscopic closure of a recurrent tracheoesophageal fistula with fibrin glue in a child. Int J Pediatr Otorhinolaryngol 1999;48(3):259-63.
30. Willetts IE, Dudley NE, Tam PK. Endoscopic treatment of recurrent tracheo-oesophageal fistulae: long-term results. Pediatr Surg Int 1998; 13(4):256-8.
31. Montedonico S, Diez-Pardo JA, Lassaletta L, et al. [Tissue adhesives in closing of fistulas after surgery of esophageal atresia]. Cir Pediatr 1999;12(3):110-2.
32. Wiseman NE. Endoscopic closure of recurrent tracheoesophageal fistula using Tisseel. J Pediatr Surg 1995;30(8):1236-7.
33. Richter GT, Ryckman F, Brown RL, et al. Endoscopic management of recurrent tracheoesophageal fistula. J Pediatr Surg 2008;43(1):238-45.
34. Quan L, Smith DW. The VATER association. Vertebral defects, anal atresia, T-E fistula with esophageal atresia, radial and renal dysplasia: a spectrum of associated defects. J Pediatr 1973;82(1):104-7.
35. Rabinowitz JG, Moseley JE, Mitty HA, et al. Trisomy 18, esophageal atresia, anomalies of the radius, and congenital hypoplastic thrombocytopenia. Radiology 1967;89(3):488-91.
36. Spitz L. Esophageal atresia and tracheoesophageal fistula in children. Curr Opin Pediatr 1993;5(3):347-52.
37. Couriel JM, Hibbert M, Olinsky A, et al. Long term pulmonary consequences of oesophageal atresia with tracheo-oesophageal fistula. Acta Paediatr Scand 1982;71(6):973-8.
38. Benjamin B. Tracheomalacia in infants and children. Ann Otol Rhinol Laryngol 1984;93(5):438-42.
39. Corbally MT, Spitz L, Kiely E, et al. Aortopexy for tracheomalacia in oesophageal anomalies. Eur J Pediatr Surg 1993;3(5):264-6.
40. Malone PS, Kiely EM. Role of aortopexy in the management of primary tracheomalacia and tracheobronchomalacia. Arch Dis Child 1990;65(4):438-40.
41. LeSouef PN, Myers NA, Landau LI. Etiologic factors in long-term respiratory function abnormalities following esophageal atresia repair. J Pediatr Surg 1987;22(10):918-22.
42. Halsband H. Esophagus replacement by free, autologous jejunal mucosa transplantation in long-gap esophageal atresia. Prog Pediatr Surg 1986;19:22-36.
43. Moutsouris C, Barouchas G, Karayannacos P, et al. Tubular musculopleural pedicle grafting of esophageal long gaps in dogs. J Pediatr Surg 1987;22(2):117-9.
44. Levine JJ, et al. Manometric variations following spiral myotomy for long-gap esophageal atresia. J Pediatr Gastroenterol Nutr 1990;10(3): 380-4.
45. Shoshany G, Kimura K, Jaume J, et al. A staged approach to long gap esophageal atresia employing a spiral myotomy and delayed reconstruction of the esophagus: an experimental study. J Pediatr Surg 1988;23(12):1218-21.
46. Shono T, Suita S. Motility studies of the esophagus in a case of esophageal atresia before primary anastomosis and in experimental models. Eur J Pediatr Surg 1997;7(3):138-42.
47. Spitz L, Kiely E, Pierro A. Gastric transposition in children: a 21-year experience. J Pediatr Surg 2004;39(3):276-81, discussion 276-81.
48. Tannuri U, Tannuri AC, Goncalves ME, et al. Total gastric transposition is better than partial gastric tube esophagoplasty for esophageal replacement in children. Dis Esophagus 2008;21(1):73-7.
49. Ein SH. Gastric tubes in children with caustic esophageal injury: a 32-year review. J Pediatr Surg 1998;33(9):1363-5.
50. Hamza AF, Abdelhay S, Sherif H, et al. Caustic esophageal strictures in children: 30 years' experience. J Pediatr Surg 2003;38(6):828-33.
51. Tannuri U, Maksoud-Filho JG, Tannuri AC, et al. Which is better for esophageal substitution in children, esophagocoloplasty or gastric transposition? A 27-year experience of a single center. J Pediatr Surg 2007;42(3):500-4.
52. Ring WS, Varco WL, L'Heureux PR, et al. Esophageal replacement with jejunum in children: an 18 to 33 year follow-up. J Thorac Cardiovasc Surg 1982;83(6):918-27.
53. Bax NM, van der Zee DC. Jejunal pedicle grafts for reconstruction of the esophagus in children. J Pediatr Surg 2007;42(2):363-9.
54. Saeki M, Tsuchida Y, Ogata T, et al. Long-term results of jejunal replacement of the esophagus. J Pediatr Surg 1988;23(5):483-9.
55. Jaureguizar E, Vasquez L, Murcia J, et al. Morbid musculoskeletal sequelae of thoracotomy for tracheoesophageal fistula. J Pediatr Surg 1985;20(5):511-4.
56. Chetcuti P, Myers NA, Phelan PD, et al. Chest wall deformity in patients with repaired esophageal atresia. J Pediatr Surg 1989;24(3): 244-7.
57. Taguchi T. Current progress in neonatal surgery. Surg Today 2008; 38(5):379-89.
58. Bianchi A, Sowande O, Alizai NK, et al. Aesthetics and lateral thoracotomy in the neonate. J Pediatr Surg 1998;33(12):1798-800.
59. Koivusalo A, Pakarinen MP, Turunen P, et al. Health-related quality of life in adult patients with esophageal atresia: a questionnaire study. J Pediatr Surg 2005;40(2):307-12.
60. Deurloo JA, Ekkelkamp S, Hartman EE, et al. Quality of life in adult survivors of correction of esophageal atresia. Arch Surg 2005;140(10): 976-80.