Esophageal Atresia and Tracheoesophageal Fistula

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8.1 Introduction

The treatment of esophageal atresia and tracheoesophageal fistula, although still a challenge, represents one of the true successes of newborn surgery. Thomas Gibson first described the classic form of esophageal atresia with tracheoesophageal fistula in 1697 [1]. In 1888, Charles Steele attempted the first surgical repair of a pure esophageal atresia. He carried out a gastrotomy and attempted to pass a steel probe through the suspected esophageal membrane. At autopsy, the probe exited the lower esophageal pouch but the two ends of the esophagus were noted to end blindly with a gap between them of 1.5 inches with no connection that could be identified [2]. Attempted repair of esophageal atresia with a tracheoesophageal fistula employing fistula division and primary anastomosis was first reported by Robert Shaw in Dallas in 1938 [3] but first attempted by Thomas Lanman in Boston in 1936, who later reported it along with 4 other attempts and 27 additional cases of esophageal atresia in 1940 [4]. All of these attempts were unsuccessful, but not all because of technical issues. After 5 failed attempts, the first in 1939, Cameron Haight undertook the first successful primary repair of esophageal atresia in 1941 using a left extrapleural approach, fistula ligation, and a single-layer anastomosis [5]. Over the years, improvements in surgical technique and neonatal care have improved outcomes in the treatment of esophageal atresia and tracheoesophageal fistula. In 1994, Spitz et al. created a system to stratify outcome based on birth weight (>1,500 g or <1,500 g) and the presence of a major cardiac anomaly [6]. In 2006, Spitz compared results in the present era to the 1994 results and noted that survival of babies with birth weight >1500 g and no cardiac anomalies was similar: 97% in 1994 *versus* 98.5% in 2006. Survival for babies that weighed <1,500 g at birth, or had a cardiac anomaly, improved from 59% to 82% during that interval. The babies with the worst outcomes, those that weighed <1,500 g at birth and had a cardiac anomaly, improved from 22% in the early era to 50% [7]. In 2009, Okamoto et al. reviewed the Japanese experience, and proposed a modification to the Spitz classification depicted in Table 8.1 [8].

8.2 Embryology

The trachea and esophagus develop from the primitive foregut during the fourth week of human embryonic development. The ventral

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Table 8.1 Survival of infants with esophageal Atresia with or without tracheoesophageal fistula based on birth weight and cardiac anomalies

Class	Birth weight (g)	Major cardiac anomaly	Survival (%)
Low risk	>2,000	No	100
Moderate risk	<2,000	No	82
Relatively high risk	>2,000	Yes	72
High risk	<2,000	Yes	27

portion of the foregut gives rise to the trachea and lungs, whereas the esophagus develops from the dorsal aspect [9]. Several authors have attempted to explain this separation, but the exact mechanism remains unknown [10]. The development of esophageal atresia and tracheoesophageal fistula appears to be a complex multifactorial process involving genetic and environmental factors to some degree. Up to 10% of patients with esophageal atresia have a defined genetic syndrome that can be diagnosed, leaving the remaining 90% with an unknown etiology for their malformation [11]. The environmental factors that have been suggested include: exposure to methimazole [12], diethylstilbestrol [13] or exogenous sex hormones [14]; infectious diseases [15]; combination of use of alcohol and tobacco by the mother [16]; maternal employment in agriculture or horticulture [17]; first-trimester maternal diabetes mellitus [18]; advanced maternal age [19]. The genetic syndromes associated with esophageal atresia and tracheoesophageal fistula include trisomies as well as single gene disorders. The trisomies include Down syndrome (21), Edwards syndrome (18), and Patau syndrome (13) [20].

The single gene disorders include CHARGE syndrome (CHD7 gene) [21], DiGeorge syndrome (TBX1 gene) [22], Feingold syndrome (MYCN gene) [23], Opitz syndrome (MID1 gene) [24], AEG syndrome (SOX2 gene) [25], and Fanconi anemia (FANCA, FANCC, and FANCG genes) [26]. Despite many recent advances, there remains a substantial gap in our understanding of the environmental, genetic, and other factors leading to abnormal development of the foregut.

8.3 Epidemiology

Esophageal atresia and tracheoesophageal fistula occurs in 1 out of every 3,500 live births [20]. The incidence varies depending on geographic location: 1 in 2,440 in Finland [27], 1 in 3,300 in England [28], and 1 in 4,500 in Australia [29] and the USA [30]. The risk that a second child will be born with esophageal atresia and tracheoesophageal fistula in the same family is approximately 1%. If one twin is born with esophageal atresia and tracheoesophageal fistula, there is only a 2.5% chance that the second twin will also have the anomaly [31]. This suggests that hereditary factors play a minimal part in the etiology of esophageal atresia. Parental characteristics that increase the risk of having a child with esophageal atresia and tracheoesophageal fistula include low maternal parity, older maternal age [32], older paternal age [33], and being a Caucasian mother compared with being a mother from other ethnic groups [32]. Obesity and low socioeconomic status of the mother are not risk factors [34].

8.4 Associated Anomalies

Although the surgical repair of esophageal atresia and tracheoesophageal fistula requires technical expertise, the outcome depends on the degree of prematurity and the presence of associated anomalies (especially cardiac and chromosomal abnormalities). Children born with esophageal atresia have a higher incidence of prematurity than the general population, most likely due to the polyhydramnios

caused by fetal esophageal obstruction [35]. Esophageal atresia and tracheoesophageal fistula present in two forms, either in isolation (50%) or as a syndromic form (50%) associated with other abnormalities [36]. Half of those in the syndromic group have random associated anomalies whereas the other half could be classified into a recognizable malformation syndrome or pattern. These syndromes or patterns include chromosomal abnormalities (trisomies 13, 18, and 21) occurring in 8% of esophageal atresia births, and several syndromes and associations, the most frequent being the VACTERL association. The VACTERL association occurs in 10–19% of cases. It consists of a non-random association of defects without specific features including **ve**rtebral, **a**norectal, **c**ardiac, **t**racheal, **e**sophageal, **r**enal, and **l**imb abnormalities. The presence of ≥3 of these abnormalities constitutes the VACTERL association [37]. The remaining syndromes and associations include the CHARGE, SCHISIS, Potter's, Feingold, Roger's, Opitz G, and Goldenhar's syndromes, and Fanconi anemia [36]. Combining data from 12 observational studies that involved 5,242 children with esophageal atresia and tracheoesophageal fistula, the following distribution of associated anomalies were found: cardiac 20.3%, digestive 16.0%, musculoskeletal 15.1%, urogenital 13.6%, central nervous system (CNS) 5.3%, and pulmonary 4.3% [37–48]. Following the initial repair or gastrostomy tube placement, an infant may develop persistent emesis. Infants with esophageal atresia and tracheoesophageal fistula have a higher incidence of pyloric stenosis than expected in the normal population [49].

8.5 Anatomy

An understanding of the anatomy involved with each case of esophageal atresia and tracheoesophageal fistula is important when devising a treatment strategy. There have been several classification systems, but a description of each type is the easiest and most practical way to depict the five combinations of esophageal atresia and tracheoesophageal fistula (Fig. 8.1). The most common configuration is esophageal atresia with a distal tracheoesophageal fistula. This configuration occurs in 86% of cases [50]. The proximal esophagus ends blindly in the upper mediastinum. The distal esophagus is connected to the tracheobronchial tree usually just above or at the carina. The second most common type is the isolated esophageal atresia without a tracheoesophageal fistula. This configuration occurs in 8% of cases [50]. The proximal esophagus ends blindly in the upper mediastinum, and the distal esophagus is also blind-ending and protrudes varying distances above the diaphragm. The distance between the two ends is often too far to bring together initially. The third most common configuration, occurring in 4% of cases [50], is a tracheoesophageal fistula without esophageal atresia. The esophagus extends in continuity to the stomach, but there is a fistula between the esophagus and trachea. The fistula is usually located in the upper mediastinum, running from a proximal orifice in the trachea to a more distal orifice in the esophagus. This is also known as an "H" type or "N" type tracheoesophageal fistula. The remaining two forms each occur in about 1% of cases [50].

These are esophageal atresia with a proximal and distal tracheoesophageal fistula, and esophageal atresia with a proximal tracheoesophageal fistula. These two forms correspond to the initial two forms described with the addition of a proximal fistula between the upper esophageal pouch and the trachea. A proximal fistula is often difficult to diagnose preoperatively even when bronchoscopy is carried out. Because of the difficulty identifying a proximal fistula, some "recurrent" fistulas in the past may have in fact been missed proximal fistulas [51]. Again the esophageal ateasia with proximal tracheoesophageal fistula, similar to its counterpart without a proximal fistula, will have a long gap between the two ends of the esophagus, making it difficult to repair initially after birth.

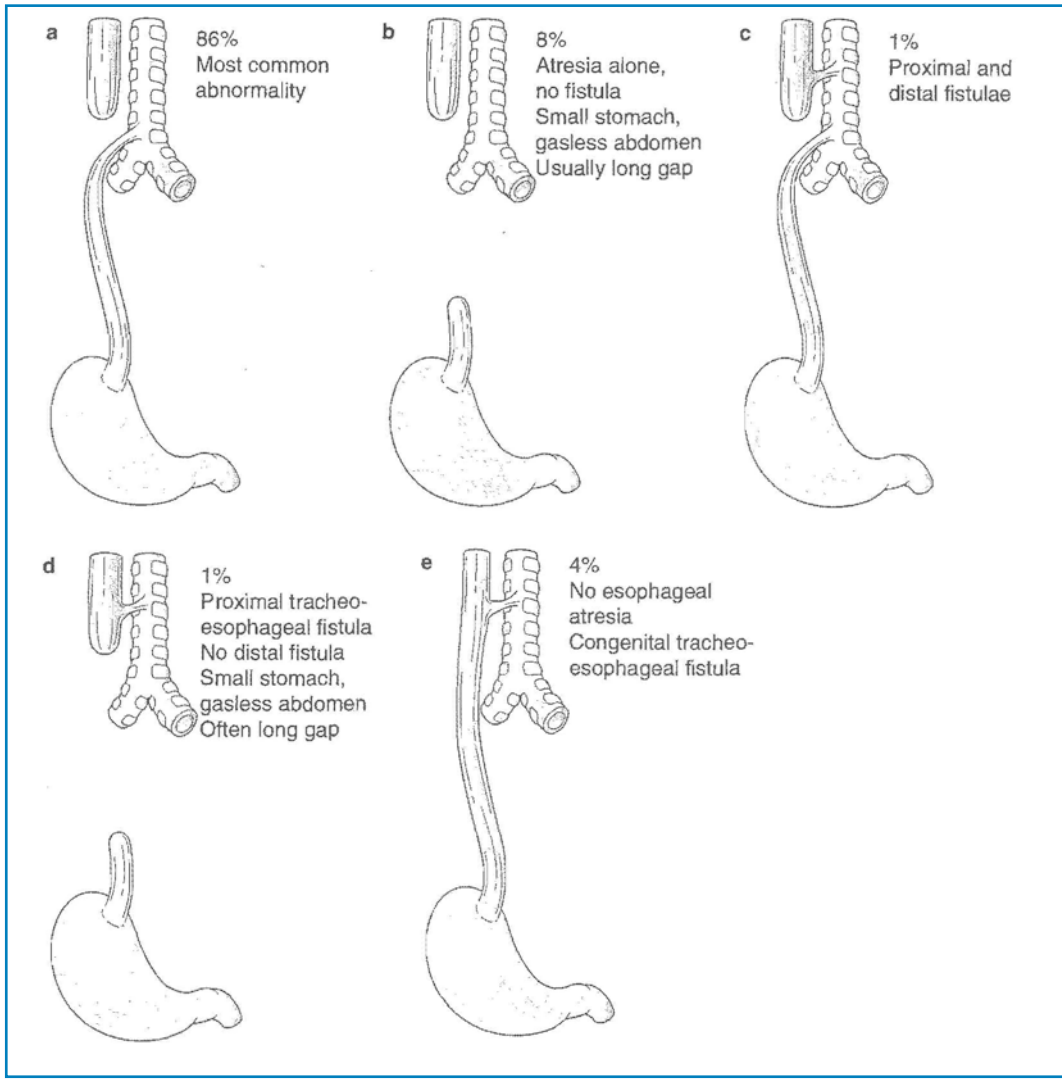

Fig. 8.1 Types of esophageal atresia and tracheoesophageal fistula

8.6 Presentation and Diagnosis

Prenatal ultrasonography attempts to predict the presence of esophageal atresia using the combination of polyhydramnios, an absent or small stomach bubble, and an "upper pouch" sign. These are non-specific findings that correctly predict esophageal atresia 56% of the time [52]. Using special image acquisition, magnetic resonance imaging (MRI) accurately demonstrates the upper pouch in fetuses with esophageal atresia [53]. Infants born with esophageal atresia and tracheoesophageal fistula have difficulty handling their oral secretions and will choke, cough, and possibly become cyanotic with their first feeding. After a feeding attempt, they will spit-up undigested formula or breast milk. This leads to an attempt to place a tube in the stomach, which does not travel as far as expected and meets resistance. A radiograph of the chest and abdomen shows the tube coiled in the esophageal pouch in the upper mediastinum, confirming esophageal atresia. The abdominal gas pattern determines if there is a distal tra-

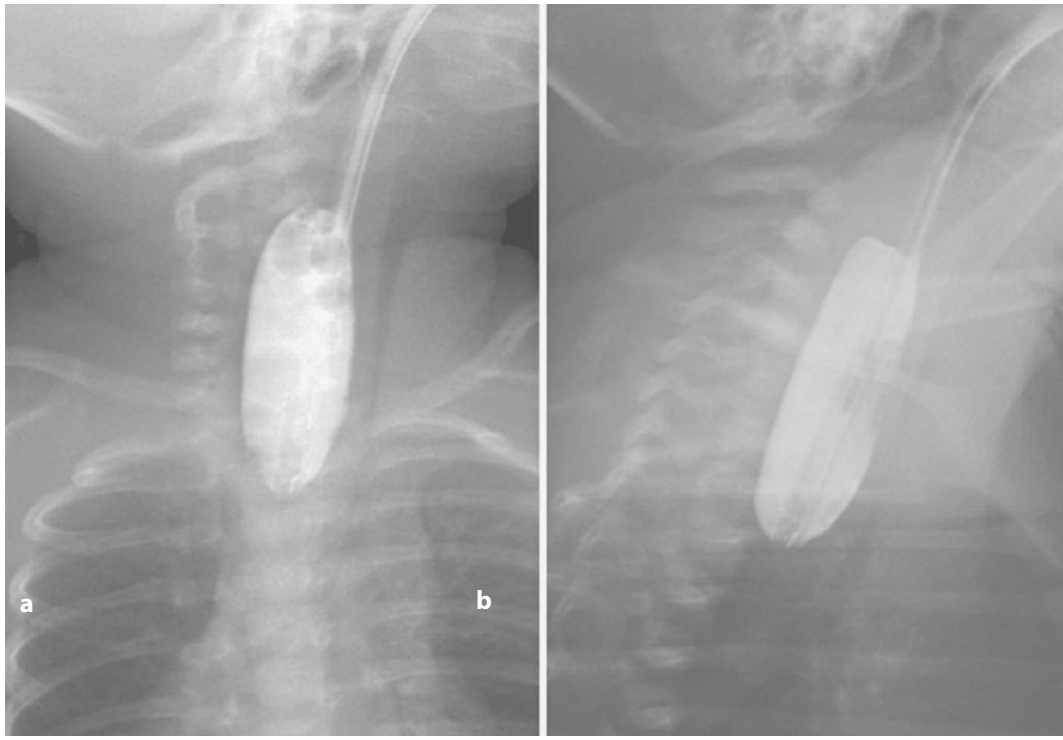

Fig. 8.2 Pouchogram obtained on a neonate without a proximal fistula. Anteroposterior view (**a**). Lateral view (**b**)

cheoesophageal fistula (gas throughout the intestines) or a pure esophageal atresia with no connection to the tracheobronchial tree (gasless abdomen). The diagnosis is made and the remainder of the preoperative evaluation attempts to define a proximal fistula between the esophageal pouch and the trachea, and to characterize associated anomalies. Chromosomal abnormalities are found with a karyotype. Four simple evaluations identify the anomalies of the VACTERL association. First, the physical examination evaluates anorectal and limb abnormalities. Second, the radiograph obtained to evaluate the placement of the esophageal tube is used to look for vertebral and limb abnormalities. Third, ultrasound of the abdomen will evaluate renal abnormalities. The pelvis should also be imaged to evaluate for a tethered spinal cord or pre-sacral mass. Finally, echocardiography evaluates cardiac anomalies and the position of the arch of the aorta (which is important in surgical planning). If a right-sided aortic arch is identified, magnetic resonance (MR) angiography is used to look for a vascular ring. A complete vascular ring is found 37% of the time in this situation [54].

Three options exist to find proximal fistulas that are not mutually exclusive. A contrast evaluation of the proximal pouch (Fig. 8.2) carried out by an experienced radiologist using a small amount of non-ionic contrast under fluoroscopic guidance often shows a proximal fistula if present. Rigid bronchoscopy just prior to surgical repair looking for an opening in the proximal membranous trachea will identify a proximal fistula. Bronchoscopy is also useful to identify the location of a distal fistula if one is present, and a laryngeotracheoesophageal cleft should be looked for as the bronchoscope is removed. The final strategy is to look for a fistula during the proximal pouch dissection. It is usually encountered near the distal aspect of the pouch, but can be located up in the neck [55]. A clue that a proximal fistula is present is that the proximal pouch will not be as dilated or thick-walled as expected because the fistula relieves

the distending pressure in the proximal pouch pre-natally and post-natally.

A tracheosophageal fistula without esophageal atresia (H-type fistula) may not present in the initial neonatal period and can be more difficult to diagnose. A tube will go into the stomach if passed, but persistent coughing and choking with oral feeds should prompt a search for an isolated fistula. A prone pull-back esophagram or bronchoscopy with esophagoscopy using methylene blue as a dye is used to find an isolated fistula. Placement of a catheter across the fistula during bronchoscopy aids the dissection greatly.

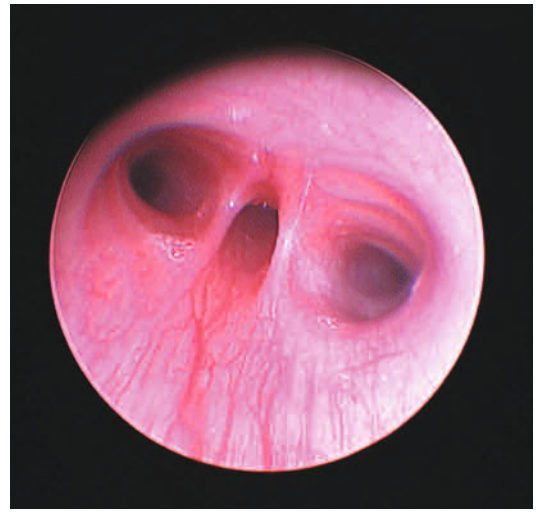

Fig. 8.3 Rigid bronchoscopic view of the carina with a distal tracheoesophageal fistula in the center

8.7 Treatment

After making the diagnosis, appropriate plans for repair should be made. In healthy newborns, the repair can be done in the first 24 h of life to minimize the risk of aspiration and the resulting pneumonitis. Prior to the procedure, the infant should be positioned supine with the head of the bed elevated 30–45°. A tube should be in the proximal pouch to constantly suction saliva and prevent aspiration. Intravenous access is established for fluid instillation along with broad-spectrum intravenous antibiotics and vitamin K. The goal of surgical therapy is to divide and close the fistula between the trachea and esophagus, and return continuity to the esophagus. In most situations, a primary repair is feasible. However, special situations require different tactics (see below).

In the usual situation, a stable infant (from the hemodynamic and pulmonary standpoint) is brought to the operating room and placed under general anesthesia. Rigid bronchoscopy (Fig. 8.3) begins the procedure to locate the distal fistula, usually at or near the carina, and to look for a proximal fistula in the more proximal trachea. Although a right posterolateral thoracotomy is the standard approach for repair of esophageal atresia, preoperative echocardiography may document a right-sided aortic arch in 2% of cases, and a left posterolateral thoracotomy should be used in these cases [56]. The difficulty repairing the esophagus through the right chest with a right-sided arch is the tension required to stretch the repair over the aortic arch, leading to a high prevalence of anastomotic leaks (about 40%) [57]. A right-sided aortic arch may be discovered intraoperatively because preoperative echocardiograpy identifies as low as 20%, and up to only 62% of the right-sided arches correctly [54, 57]. In that situation, the repair is attempted through the right chest, and if it cannot be completed, the tracheoesophageal fistula is divided, the right chest closed, and a left thoracotomy used to complete esophageal repair.

For the infant with the typical left-sided aortic arch, a right-sided posterolateral thoracotomy using a muscle-splitting, retropleural approach gives access to the mediastinal structures. The azygos vein is divided, revealing the tracheoesophageal connection. The distal esophagus is divided at the level of the trachea and the defect in the trachea closed (Fig. 8.4). Dissection of the distal esophagus is minimized to protect its segmental blood supply. The thyrocervical trunk provides a rich blood supply to the proximal esophagus, allowing extensive dissection. The anesthesiologist pushes on the tube in the proximal pouch to help identify it in the upper mediastinum. Dis-

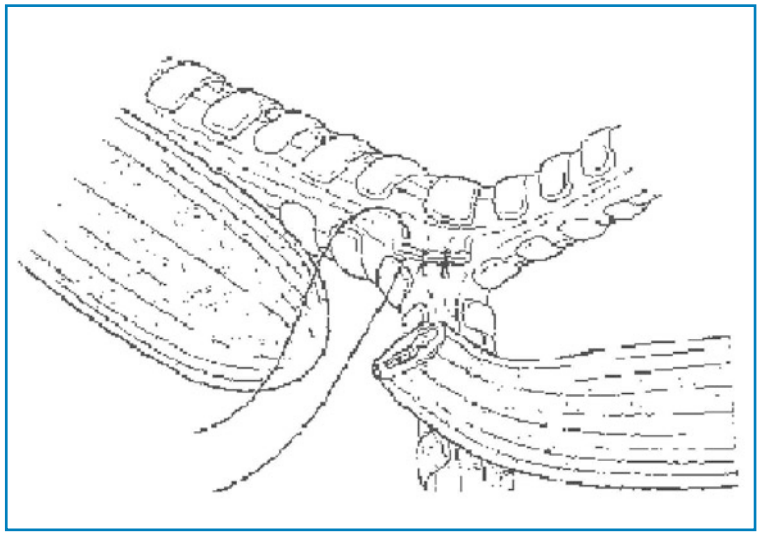

Fig. 8.4 Closure of the trachea after division of a distal tracheoesohageal fistula

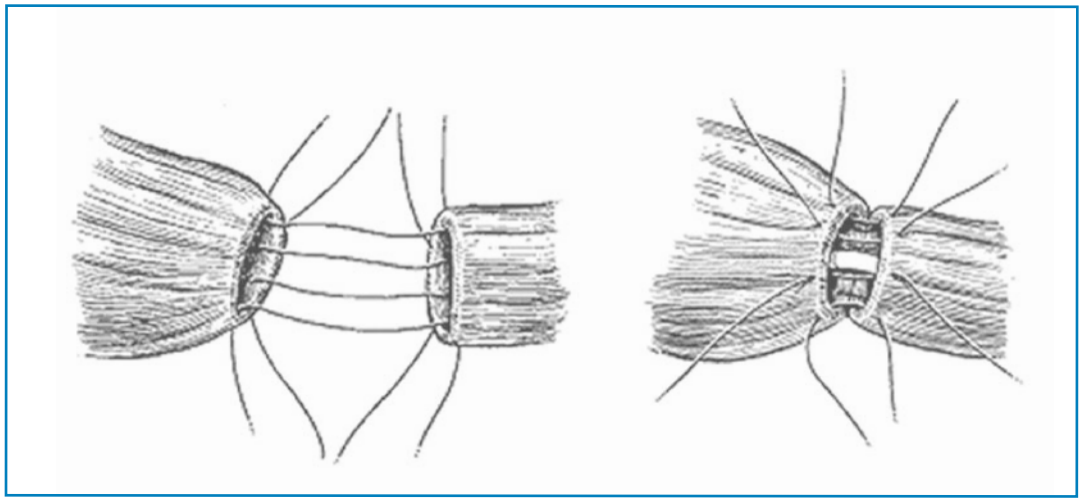

Fig. 8.5 Esophageal anastomosis with a nasogastric tube used as a stent

section of the proximal pouch proceeds on the thickened esophageal wall to avoid tracheal injury, and continues as high as possible to gain length for a tension-free anastomosis, and to search for the rare proximal fistula. A single-layered end-to-end interrupted anastomosis brings the ends of the esophagus together (Fig. 8.5). A nasogastric tube is passed through the anastomosis to ensure patency of the distal esophagus, and allow initial gastric decompression and eventual enteral feeds. A chest tube is placed in the retropleural space to control a potential leak. Some surgeons opt not to use a chest tube if the pleura remains intact. One advantage of the retropeural approach is that if the anastomosis leaks it will not soil the hemithorax and result in an empyema. A leak into the retropleural space will result in an esophagocutaneous fistula that will almost always close spontaneously.

Recently, thoracoscopic repair of esophageal atresia and tracheoesphageal fistula has proven feasible with similar outcomes to those seen with open repair [58]. Several large series

have been reported, including a multicenter series with 103 thoracoscopic repairs [59] and a single-institution series of 51 thoracoscopic repairs [60]. The left semi-prone position allows the lung to fall anterior and, along with CO_2 insufflation with 5 mmHg, provides an excellent view of the posterior mediastinum. Three or four trocars are used to disconnect the fistula and carry out the esophageal anastomosis. The purported benefits of the thoracoscopic approach include less pain from the incisions, less shoulder asymmetry and scoliosis, better cosmetic effect, and a magnified surgical view. The thoracoscopic approach requires advanced endosurgical skills because it is technically demanding.

After surgery (whether open or thoracoscopic) the infant returns to the Intensive Care Unit (ICU) and continues on intravenous nutrition and antibiotics. Special nursing care with frequent oropharyngeal suctioning and positioning with the head of the bed elevated 30–45° helps to prevent aspiration. Feedings may be started through a trans-anastomotic tube 2–3 days after surgery. Acid-suppressive therapy prevents irritation of the anastomosis and subsequent stricture formation. On postoperative days 5–7 esophagography can be used to check the integrity of the anastomosis. If no leak is seen, feedings are started orally, and the chest tube removed the following day. If a leak is present, conservative treatment with intravenous nutrition, broad-spectrum antibiotics, and chest-tube drainage continues. Another esophagram is ordered for the following week. These leaks will invariably close without further surgical intervention [61]. Only complete disruption of the anastomosis requires further intervention. A cervical esophagostomy drains the proximal pouch, the distal esophagus is tied off, and the mediastinum is drained widely.

8.8 Special Situations

Most cases of esophageal atresia and tracheoesophageal fistula can be handled routinely, but three situations require further discussion, i.e., infants with: severe respiratory disease in which the tracheoesophageal fistula contributes to ventilator insufficiency; long-gap esophageal atresia; an H-type tracheoesophageal fistula.

Infants with respiratory insufficiency and a tracheoesophageal fistula are often premature with lung immaturity and require a significant amount of ventilatory support. Breaths provided by the ventilator preferentially enter the distal esophagus through the fistula due to the high resistance in the airways produced by the non-compliant lungs. This allows a significant portion of each breath to enter the abdomen through the fistula and distal esophagus, producing abdominal distension and elevation of the hemidiaphragm, leading to further respiratory compromise. There are several options in this situation. High-frequency ventilation decreases the portion of the tidal volume lost through the fistula. Advancing the endotracheal tube distal to the fistula opening prevents further loss of ventilation, but is not always possible, and can be dangerous if the fistula is intubated [62]. A Fogarty catheter placed in the fistula bronchoscopically and inflated temporarily occludes the fistula but has a tendency to dislodge [63]. The Fogarty catheter may also be placed through a gastrostomy tube into the distal esophagus, or through the fistula into the trachea to prevent further runoff. The Fogarty catheter can then be secured well to the gastrostomy tube [64]. When a gastrostomy tube is present, it can be connected to a pleurovac and placed in an underwater seal to increase the resistance of the tract and reduce airflow through the fistula [50]. If these techniques fail, these infants require an urgent thoracotomy to ligate the tracheoesophageal fistula, prevent further respiratory decompensation, and ameliorate the risk of gastric perforation. Often the infant's respiratory status stabilizes in the operating room after fistula ligation, allowing completion of the repair [65]. If the infant remains unstable, the distal esophagus is secured to the prevertebral fascia, the chest closed, a gastros-

tomy tube placed (if not already present) and definitive repair delayed until the respiratory status stabilizes.

A long gap between the two esophageal segments comprises the next special situation. This often occurs with pure esophageal atresia or esophageal atresia with a proximal fistula. Both of these present with a radiographic picture of a gasless abdomen. On occasion, an infant with esophageal atresia and a distal tracheoesophageal fistula may have a long gap between the two esophageal segments. Initially, the infant requires a gastrostomy tube for enteral feedings while waiting for the two esophageal segments to grow adequately to allow a primary anastomosis. The stomach is quite small in these infants because it was unused during fetal life and has not yet stretched to its full capacity. During gastrostomy placement, care must be taken to avoid injury to the small stomach and its blood supply along the greater curvature. Careful placement will allow for future use of the stomach for an esophageal replacement if necessary. During placement of the gastrostomy tube, an estimation of the distance between the esophageal segments is obtained. Several methods utilizing metal sounds, dilators, contrast and endoscopy exist. The most accurate method uses fluoroscopy to measure the distance between a dilator placed in the upper esophageal pouch and an endoscope placed through the gastrostomy site and driven into the blind distal esophageal pouch [66]. If the ends of the esophageal segments are >3 vertebral-bodies apart they will not be easily connected. The infant should be fed *via* the gastrostomy tube with the head of the bed elevated 30–45° and a tube in the proximal pouch to control the secretions. During the first several months of life, the gap between the two segments of the esophagus shortens due to spontaneous growth of the atretic esophagus [67]. Stretching of the upper and (less frequently) the lower pouch twice a day by bougienage may help bring the two ends together [68]. The distance between the proximal and distal segments of the esophagus is measured every 2–4 weeks and, if they are within 2–3 vertebral bodies (which occurs in about 70% of infants [69]), a thoracotomy and attempted anastomosis are done.

There are several techniques that can be used to gain length on the esophageal segments during the procedure. These include complete dissection of the upper pouch to the thoracic inlet. A circular myotomy of Livadit is carried out on the upper pouch and produces approximately 1 cm of length for each myotomy [70]. Use of a circular myotomy is shown in Fig. 8.6. A tubularization graft of the upper pouch can be created and connected to the distal esophagus (Fig. 8.7) [71]. The distal esophagus can also be mobilized (despite its segmental blood supply) down through the hiatus to gain length [50]. If these maneuvers fail to provide an adequate anastomosis, other options include a staged esophageal replacement with a left cervical esophagostomy, or an immediate esophageal substitution with a gastric transposition, a gastric tube, or a colon interposition to replace the native esophagus. An immediate gastric transposition is our preferred approach. If the gap remains >3 vertebral bodies and the ends of the esophageal segments are no longer approaching each other, the infant may require a cervical esophagostomy and esophageal replacement at 9–12 months of age. Waiting >4 months rarely provides extra growth of the esophageal ends resulting in primary anastomosis. The esophagostomy allows sham feeds to prevent oral aversion and subsequent feeding problems without the risk of aspiration while awaiting esophageal replacement.

Two other surgical options to maintain the native esophagus include the Foker traction technique and the extrathoracic elongation technique of Kimura. The Foker technique involves placing traction sutures on both segments of esophagus and attaching them under tension to the prevertebral fascia if the gap is of moderate length, or bringing them out through the back and increasing tension on them sequentially over the ensuing 2 weeks. When the ends of the two segments of esophagus approach each other, a repeat thoracotomy with primary anastomosis completes the repair

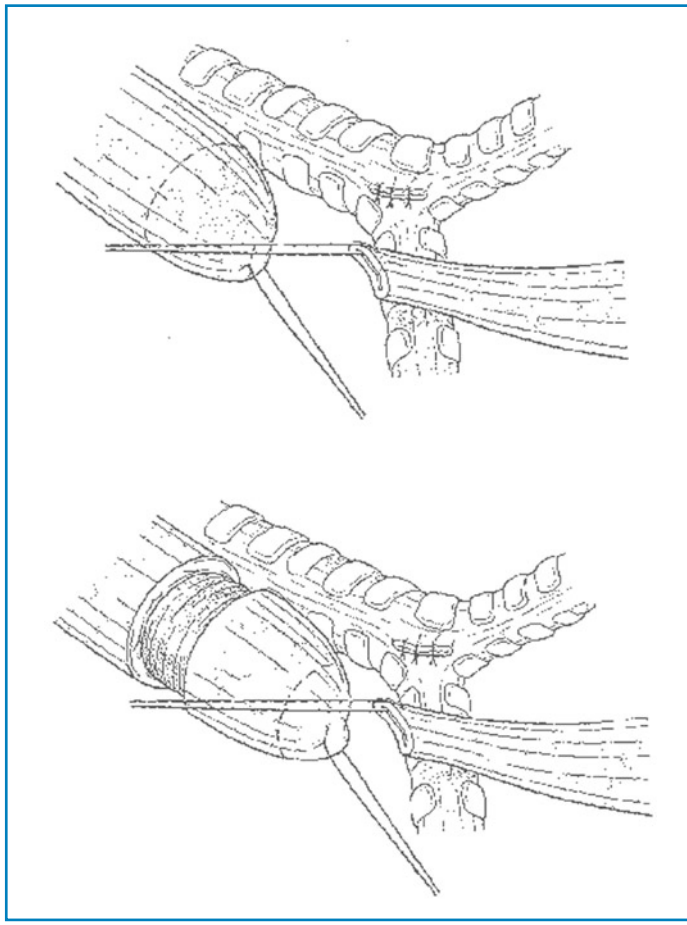

Fig. 8.6 A circular myotomy of the upper pouch extending its length by 1 cm

(Fig. 8.8) [72]. The Kimura technique involves mobilization of the proximal pouch and bringing it out as an end cervical esophagostomy. Then, every 2–3 weeks, the esophagus is mobilized and moved down the anterior chest wall until enough length is obtained to complete an end-to-end esophageal anastomosis *via* a thoracotomy [73].

The final special situation is the H-type tracheoesophageal fistula without esophageal atresia. To repair this fistula, a Fogarty catheter is placed across the fistula using rigid bronchoscopy and esophagoscopy. Exploration of the right neck through an incision just above the clavicle allows identification and division of the fistula. Palpation of the Fogarty catheter assists in finding the fistula (which is quite difficult without a catheter *in situ*). If possible, muscle or other vascularized tissue is placed between the suture lines to help prevent recurrence of the fistula.

8.9 Complications

Complications after repair of esophageal atresia and tracheoesophageal fistula relate to the anastomosis and the underlying disease. The anastomotic problems include anastomotic leaks, anastomotic strictures, and recurrent formation of a tracheoesophageal fistula. The issues related to the underlying disease include gastroesophageal reflux and tracheomalacia. The incidence of anastomotic problems seen after repair varies directly with the amount of tension used to create the anastomosis.

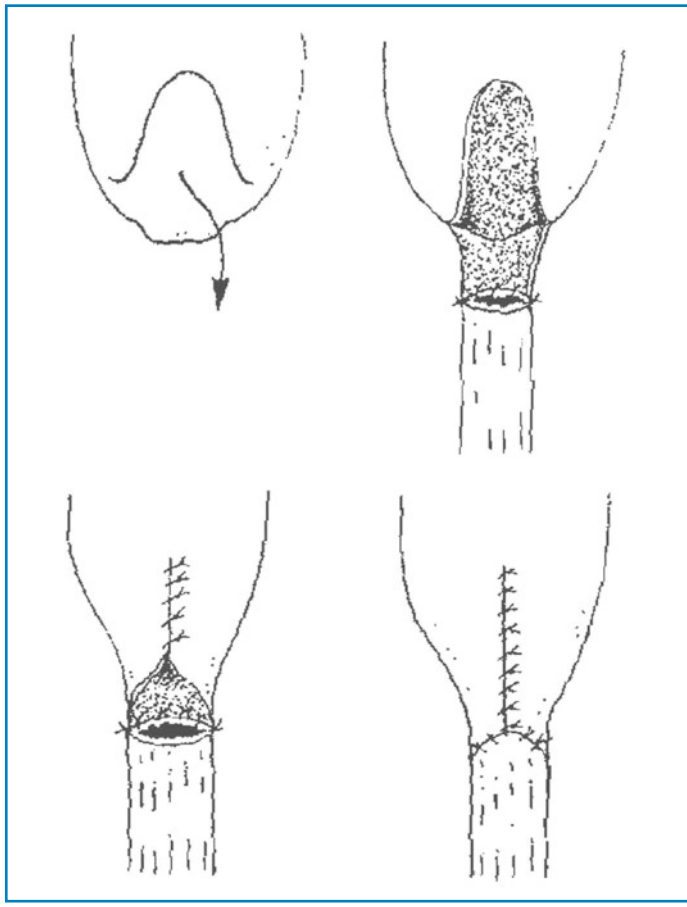

Fig. 8.7 Tubularization graft of the upper pouch

The incidence of leak at the anastomosis varies from 5% to 20% [45]. Most of these leaks seal in 1–2 weeks with conservative management, including parenteral nutrition, intravenous antibiotics, drainage, and withholding oral feeds. Complete disruption of the anastomosis, a rare complication occurring in <2% of cases, causes tension pneumothorax and significant salivary drainage from the chest tube. This scenario may require early thoracotomy and revision of the failed anastomosis, or cervical esophagostomy, gastrostomy, and subsequent esophageal replacement.

Anastomotic strictures occur in one-third to one-half of repairs [74]. Factors implicated in stricture formation include tension at the anastomosis, anastomotic leak, and gastroesophageal reflux [75, 76]. All repairs will show some degree of narrowing at the anastomosis, so dilations are reserved for symptomatic strictures, associated respiratory symptoms, foreign-body obstruction or those causing dysphagia. Most strictures require repeat dilation every 3–6 weeks over a 3–6-month period. Two recent series of esophageal strictures after repair of esophageal atresia revealed an average of 2.8–3.2 dilations required per stricture over an average of 7 months [75, 76]. Recalcitrant strictures often occur in association with gastroesophageal reflux and do not resolve until the reflux is controlled by medical or surgical means.

The prevalence of recurrent formation of tracheoesophageal fistulas ranged from <1% to 12% in various series [45, 77-80]. These children present with coughing, choking, and

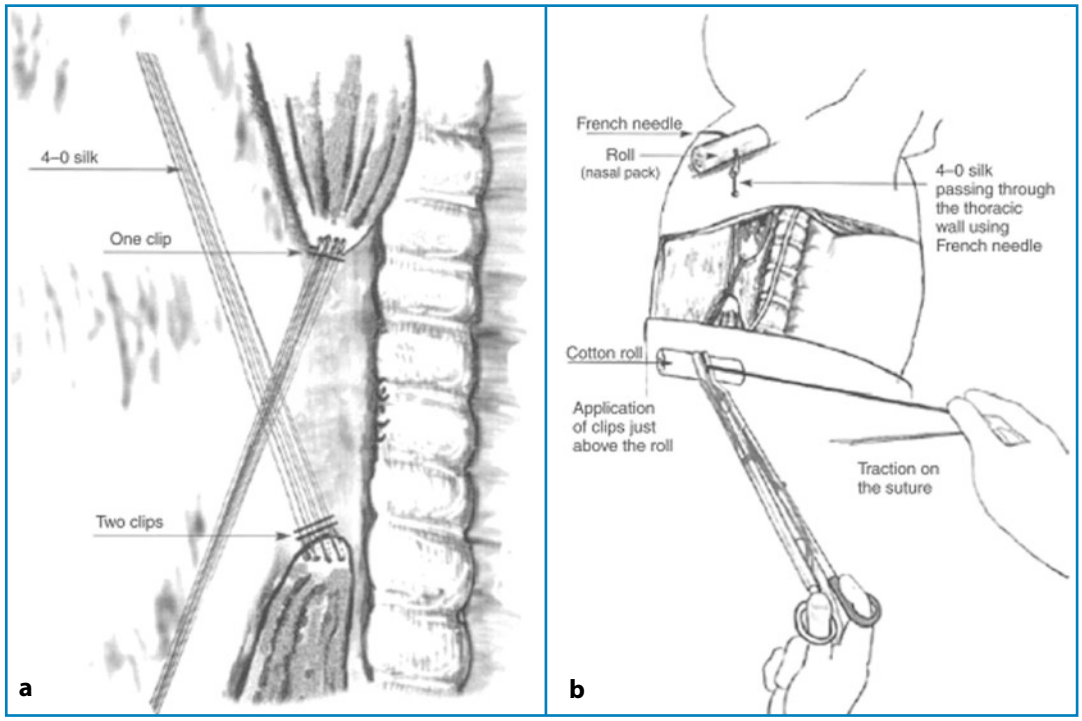

Fig. 8.8 Foker traction technique. Traction sutures are placed on each esophageal segment (**a**). The sutures are brought out the back of the infant and traction is increased incrementally until the two segments are close enough for anastamosis (**b**)

occasional cyanotic episodes with feeding, and with recurrent pulmonary infections. Recurrent fistulas are often associated with anastomotic leaks, but the possibility of a missed proximal fistula must also be entertained [81]. These fistulas are often small and sometimes difficult to diagnose. A prone, pullback esophagram combined with bronchoscopy with esophagoscopy are used to diagnose recurrent fistulas. A repeat right thoracotomy with closure of the fistula is a difficult procedure. Placement of a ureteral catheter in the fistula at bronchoscopy just before opening the chest improves the likelihood of identifying the fistula tract. After identification and division of the fistula, a viable piece of tissue (usually a vascularized muscle flap or a portion of pleura or pericardium) placed between the suture lines helps prevent recurrence of the fistula, which occurs in 11% to 20% of these repairs [79, 80]. Other techniques to close these fistulas have been attempted with varying success, including endoscopic diathermy [82], Nd:YAG [83] or KTP laser obliteration of the fistula [84], injection of sclerosing agents [85], and injection of fibrin glue [86]. If using these approaches, it is important to obliterate the mucosal lining of the fistula tract prior injecting agents to occlude the fistula. These techniques may be repeated, but if they continue to fail, surgical closure is required.

Gastroesophageal reflux commonly occurs in patients after esophageal atresia and tracheoesophageal fistula repair. Poor distal esophageal motility resulting in abnormal esophageal clearance accompanied with an altered angle of His due to the tension on the distal esophagus predispose these children to reflux. Using videomanometry with topographic analyses, Kawahara found two subgroups of patients after repair of esophageal atresia and tracheoesophageal fistula. Neither group had esophageal contractions at the anas-

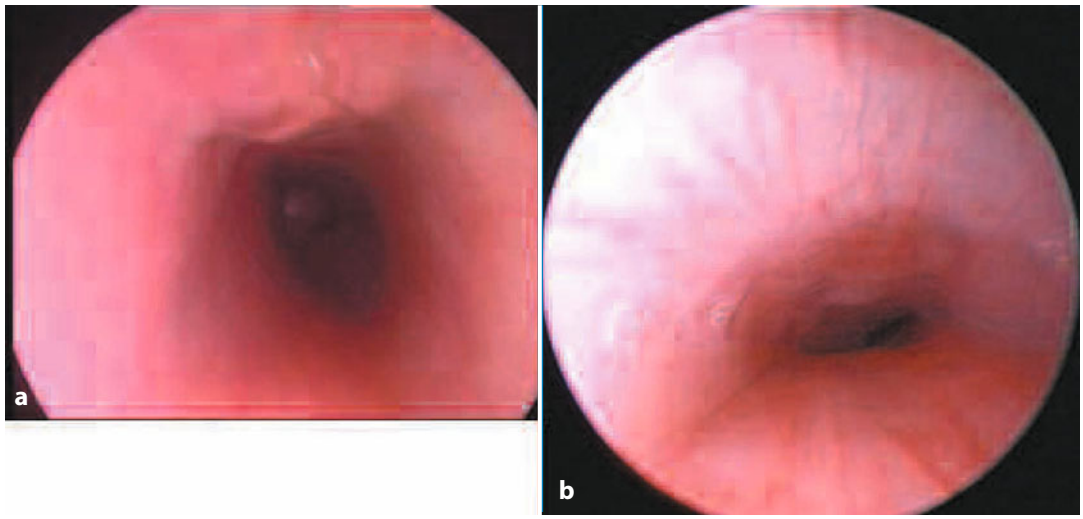

Fig. 8.9 Bronchoscopic view of the trachea in an infant with tracheomalacia. The trachea is open (**a**) during inspiration, and collapses on itself during expiration (**b**)

tomosis. One group had distal esophageal contractions and did not develop reflux, whereas the other group did not have distal contractions, and 15 of 17 developed symptomatic gastroesophagael reflux [87]. Significant gastroesophageal reflux occurs in 30% to 60% of children after repair of esophageal atresia and tracheoesophageal fistula [45, 77, 88]. In an adult series of patients born with esophageal atresia compared with normal controls, Sistonen et al. found symptomatic reflux in 34% compared with 8% of controls, dysphagia in 85% compared with 2%, and at endoscopy found more hiatial hernias (28%), strictures (8%), chronic esophagitis (8%) and Barrett's esophagus (11%) compared with controls [89]. Treatment begins initially with acid-reducing and prokinetic agents, but often patients require a fundoplication to control the reflux, especially if a stricture develops which is resistant to dilation, or if repeated pulmonary aspiration associated with reflux complicates the postoperative course. Careful consideration should be given to a partial fundoplication in these children because of their abnormal distal esophageal motility. The choice of a complete or partial fundoplication is left to the surgeon [90–92]. A comparison of fundoplications done in babies with and without esophageal atresia and tracheoesophageal fistula revealed that those with esophageal atresia had more intraoperative and postoperative complications and more problems with recurrent reflux, dysphagia and dumping after the fundoplication [93].

Significant tracheomalacia occurs in 10% to 20% of children with esophageal atresia and tracheoesophageal fistula [45, 94]. Tracheomalaciais a generalized or localized collapse of the tracheal lumen causing obstruction during respiration, and is considered pathologic if the obstruction is >50% of the lumen [95]. This results in expiratory stridor and episodes of desaturation, apnea, cyanosis, and bradycardia, and is often associated with feeds. The upper tracheal cartilage becomes weakened due to the pressure exerted from the fluid-filled dilated upper esophageal pouch during fetal life. Tracheomalacia, in its severe form sometimes prevents extubation after repair of the esophageal atresia and tracheoesophageal fistula. Determining the etiology of this symptom complex can be difficult because tracheomalacia and gastroesophageal reflux occur frequently in this population and produce similar symptoms. Rigid bronchoscopy in a spontaneously breathing patient provides the diagnosis. The trachea will flatten anteroposteriorly

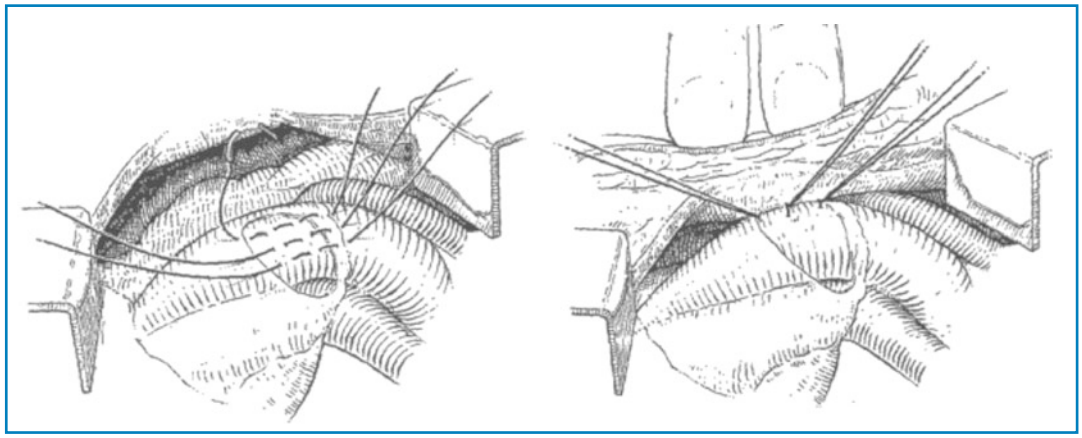

Fig. 8.10 Through an anterior second interspace thoracotomy, sutures are placed in the adventitia of the ascending aorta, fixing it to the posterior sternum, thus stenting the underlying trachea open. This is confirmed bronchoscopically during the procedure

("fishmouth") on expiration (Fig. 8.9). Tracheomalacia is often self-limiting, but may require intervention in children with severe life-threatening symptoms. If treatment with continuous positive-airway pressure is ineffective, then aortopexy [96] (Fig. 8.10) or tracheal stenting may be required [97]. Aortopexy fixes the adventitia of the aorta to the posterior sternum, thus stenting open the underlying trachea. Aortopexy remains the treatment of choice because the optimal tracheal stent for infants has not been developed [95]. Symptomatic tracheomalacia requires intervention in up to 5% of children with esophageal atresia [98].

8.10 Outcome

The outcome for infants with esophageal atresia and tracheoesophageal fistula has improved over time to the point where, unless the infant has major cardiac anomalies, significant chromosomal abnormalities, severe pulmonary complications, or a birth weight <1,500 g, he/she will survive. The long-term problems after esophageal atresia and tracheoesophageal fistula repair include pulmonary issues (especially reactive airway disease, bronchitis, and pneumonias) and upper gastrointestinal complaints of dysphagia and gastroesophageal reflux. There is a strong connection between the severity of the gastroesophageal reflux and the persistence of respiratory symptoms in these patients [99]. Pulmonary symptoms severe enough to require hospitalization occur in close to half of children after repair of their esophageal atresia and tracheoesophageal fistulas [100]. Although the pulmonary symptoms tend to persist into adulthood, they are mild and do not affect the activities of daily life [101]. The dysphagia and gastroesophageal reflux commonly seen in these children stem from the altered intrinsic innervation of the distal portion of the esophagus, leading to the dysmotility that contributes to the dysphagia and reflux. The dysmotility continues into adulthood. In manometric studies of adults with repaired esophageal atresia, the overriding long-term motility deficits are uncoordinated peristaltic activity and low-amplitude contraction of the distal esophagus. Interestingly, the swallow-induced relaxation of the lower esophageal sphincter remains normal. This abnormal esophageal motility results in dysphagia symptoms in up to 60% of adults and in gastroesophageal reflux. Symptoms of dysphagia occur more commonly in adults with pure esophageal atresia compared with those with esophageal atresia with a distal tracheoe-

sophageal fistula [102]. Using 24-h pH monitoring and esophageal biopsy data, the prevalence of gastroesophageal reflux has been documented in infants, in children up to age 10 years, and in adults after esophageal atresia repair. The prevalence of reflux was similar in the three age groups: 41% in infants, 45–50% in children up to age 10 years, and 40% in adults. In the group of children up to age 10 years, no new cases of histologic esophagitis or abnormal pH measurements occurred after age 5 years. The gastroesophageal reflux appears to develop early and persist after esophageal atresia repair [102]. Esophageal strictures are uncommon as a late complication. If a stricture does occur late in the course, it is usually associated with gastroesophagaeal reflux. Barrett's esophagus occurs in up to 11% of these patients [89]; how many of these progress on to esophageal adenocarcinoma remains uncertain [103]. Age, surgically treated anastomotic stricture in infancy, recurrent tracheoesophageal fistula, myotomy of the upper esophageal pouch, anastomotic stricture in adulthood, low distal esophageal wave amplitudes, and non-propagating peristalsis were found to be strong predictive factors for epithelial metaplasia in adults with esophageal atresia treated as infants [89]. There have been 6 reported cases of esophageal cancer in patients who had their esophageal atresia repaired at birth. Three of these were squamous cell carcinomas, and 3 were adenocarcinomas. These cases developed early between the ages of 20 years and 46 years [102]. However, a population-based, long-term follow-up study of 502 patients with repaired esophageal atresia over 50 years in Finland revealed 3 cases of cancer, none being esophageal or gastric [104]. Several quality of life (QoL) measures have been used to assess the long-term outcomes of adults after repair as an infant. A Dutch study of QoL in adults after repair of esophageal atresia and tracheoesophageal fistula compared with healthy subjects found no difference in overall physical or mental health between the two groups. However, former esophageal atresia patients reported worse "general health" and less 'vitality" than the healthy subjects because of continued gastrointestinal difficulties reported in up to one-quarter of the esophageal atresia group. Marital and family status did not differ from that of the general Dutch population [105]. A recent QoL study in patients aged 6–18 years with esophageal atresia revealed similar results: the general health was reduced compared with the reference population due to gastrointestinal symptoms and a high incidence of congenital anomalies [106]. The QoL of adults after a colonic interposition as an infant is not as good as it is for adults who had a primary repair. In addition, children with esophageal atresia and tracheoesophageal fistula have more learning, emotional, and behavioral problems than the normal population of children. This in part because of their associated anomalies, including varying degrees of prematurity, and their initial ICU course, which often involves mechanical ventilation for a period of time [107].

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