22
Congenital Malformations

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Introduction

Esophageal atresia presents some of the greatest technical hurdles that the pediatric surgeon has to face. Little more than 60 years ago esophageal atresia was considered a uniformly fatal condition. Now, survival is almost guaranteed and determined by associated abnormalities rather than the esophageal atresia itself. The focus of attention has shifted from survival to minimization of morbidity. Nevertheless, with all its variations, subtleties, technical difficulties, and the long-term implications, esophageal atresia continues to challenge the pediatric surgeon.

Historical Perspective

William Durston is believed to have provided the first description of esophageal atresia over 300 years ago in "a narrative of a monstrous birth."[1,2] After that there were several anecdotal reports until the association of esophageal atresia with other structural abnormalities was recognized by Thomas Hill in 1840.[3] In 1861 Hirschsprung brought together a series of 14 cases. A more complete account of the embryology and clinical diagnosis, including associated anomalies was provided by Morrell McKenzie in 1880.[4] Up until this time, accounts of esophageal atresia had been observational rather than interventional. The first attempt to repair esophageal atresia was by Charles Steel in 1888 when he pushed a metal probe introduced through a gastrotomy up into the lower esophageal segment while another bougie was pushed downward from above. He assumed that the esophagus was blocked by a membrane but his procedure failed and autopsy revealed a gap between the proximal and distal segments.[5] Permanent gastrostomies were performed from 1899 and it was not until the 1930s that definitive repair along the lines currently employed (i.e., division of the tracheoesophageal fistula and esophageal anastomosis) was attempted. Thomas Lanman probably performed the first definitive repair in 1936 although the first to appear in the literature was a case operated in 1938 by Robert Shaw.[6] Lanman subsequently reported that all 30 operative cases in his series died, but added that "the successful operative treatment of a patient with this anomaly is only a question of time."

The first successful primary repair of esophageal atresia was achieved by Cameron Haight in 1941. His five previous attempts at primary repair all failed.[7] By the mid to late 1940s definitive surgery for esophageal atresia was being performed throughout the world including in infants under 1.5 kg.[8] The first survivor in the Southern Hemisphere was operated in New Plymouth, a small rural hospital in New Zealand in 1948,[9] and the first Australian survivor was treated at the Royal Children's Hospital, Melbourne, the following year.[2] Over subsequent decades, deliberate staged repairs, gastrostomies, cervical esophagostomies, and chest drains all became less common. Even in the very premature infant, early primary definitive repair became routine.

By the early 1990s a few surgeons were performing thoracoscopic mobilization of the esophagus,[10] but it was not until 1999 that Rothenberg reported the first successful complete repair of esophageal atresia thoracoscopically.[11] Since then, the technique has become routine in some centers,[12,13] and has even been applied to the H-type tracheoesophageal fistula.[14]

Basic Science/Pathogenesis

Embryogenesis of Esophageal Atresia

Tracheoesophageal Separation

In general terms, the normal changes that occur during early foregut differentiation into the trachea anteriorly and esophagus posteriorly are well described, although the exact mechanism of separation of the two structures is controversial. Many morphological descriptions have been proffered over the years, each reflecting subtle differences in interpretation of sequential histological observations, microdissections, or scanning electron microscopy images. For example, evidence for an ascending tracheoesophageal septum separating the trachea from the esophagus has been conflicting.[15] Similarly, the

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description of inward growth and fusion of two lateral epithelial ridges to partition the foregut into trachea and esophagus has been disputed.[16,17] Recent techniques of computer-assisted 3D reconstruction, and recognition of the importance of apoptosis in the process of defining morphology, have aided our understanding of the mechanisms involved.

In the rat embryo, both bronchial buds appear before tracheoesophageal separation commences[18] (Fig. 22.1a). This observation is not consistent with the popular "tap water" theory proposed by O'Reilly and Muller that described how the lung bud grows caudally into the ventral foregut mesenchyme before dividing into primary bronchi[16] (Fig. 22.1b). Scanning electron microscopy and techniques of 3D reconstruction of serial sections[19] have helped clarify that tracheal separation involves three consecutive stages:

1. Formation of bronchial buds from epithelial proliferation.
2. Initiation of separation of the trachea and esophagus by epithelial apoptosis. This stage involves a clearly defined pattern of apopotosis that is extremely consistent and tightly controlled both in terms of timing and location (Fig. 22.2).
3. Completion of the separation process by epithelial proliferation[20] (Fig. 22.3).

Aberrations of the normal temporospatial characteristics of apoptosis during foregut development result in abnormal morphology[21] such as esophageal atresia and its related structural abnormalities, including tracheomalacia.

In the human the primordial tracheopulmonary anlage is detectable at 21 days after fertilization. By 32 days (8 mm) separation of the trachea and esophagus is complete. This suggests that the insult causing esophageal atresia is likely to occur between 21- and 32-days gestation.[22]

Previous theories of embryogenesis of esophageal atresia – such as pressure from embryonic cardiomegaly in association with marked dorsal curvature of the cervical region,[23] pressure from abnormal vessels derived from the caudal portion of the right dorsal aorta,[24] pressure by the pneumatoenteric recesses,[25] embryonic hyperflexion,[26] and epithelial occlusion[27] – have been disproved.

The Abnormal Notochord in Esophageal Atresia

The notochord has long been recognized as influencing axial organ development,[28,29] so it is not surprising that notochordal abnormalities are associated with malformations of the trachea and esophagus.

In rodent embryos exposed to Adriamycin that develop esophageal atresia, the notochord shows abnormal branching patterns that extend from the vertebral column into the mesenchyme. It is often close to or attached to the foregut.[19,30,31] Aberrations in the structure of the notochord correlate closely with the spectrum of abnormalities seen in the VATER association.[32] For example, an excessively ventrally placed notochord produces abnormalities of the vertebral column, especially hemivertebrae.[33] Similarly, prolonged adherence of the notochord to the foregut results in abnormal development of the mesenchyme and is associated with esophageal atresia and tracheoesophageal fistula (Fig. 22.4).[30]

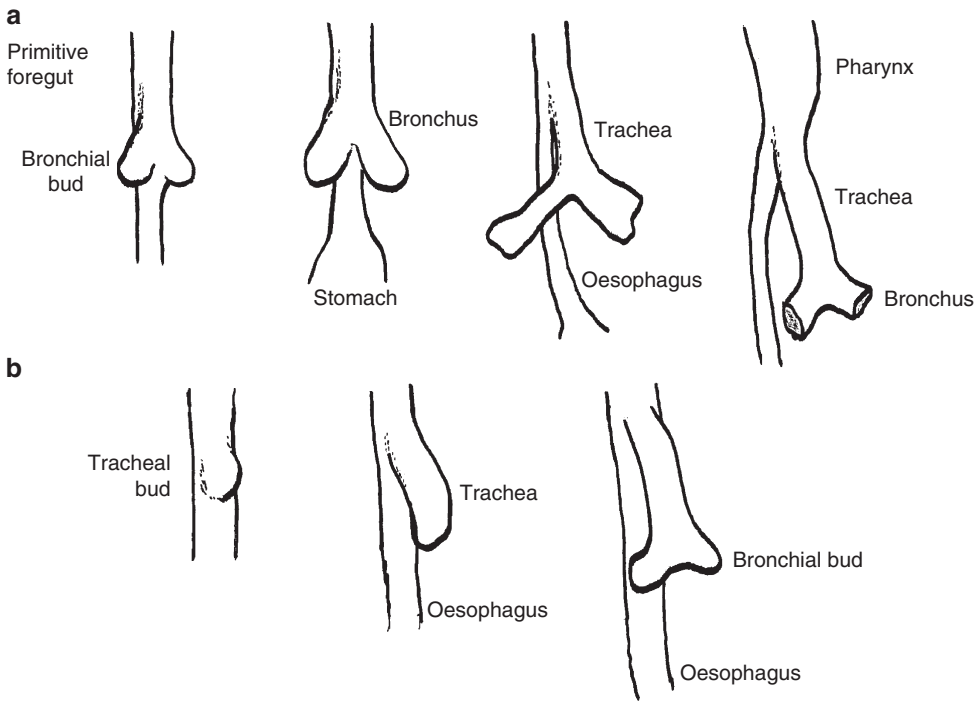


FIG. 22.1. The primitive foregut tube differentiates into trachea and esophagus. Morphological changes involve bronchial bud development before tracheoesophageal separation (**a**), contrasting with the previously-held "tapwater" theory of O'Reilly and Muller (**b**) that assumed that the trachea developed first and bronchial division occurred later

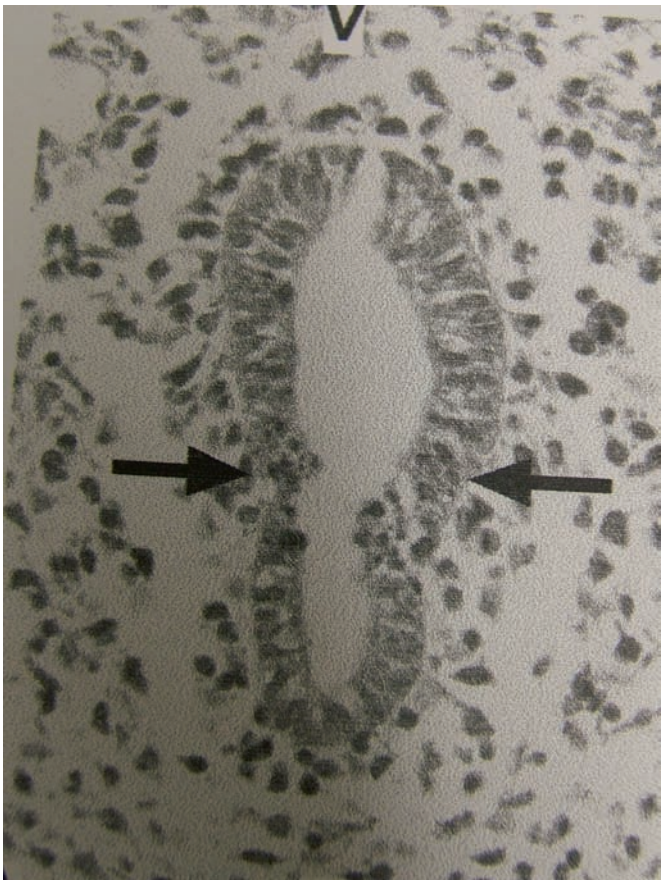


FIG. 22.2. Cross section of the primitive foregut showing apoptosis resulting in tracheoesophageal separation. Note the differences in appearance of the future respiratory epithelium (anteriorly) and the esophageal epithelium (posteriorly). The future esophageal lumen is of smaller caliber

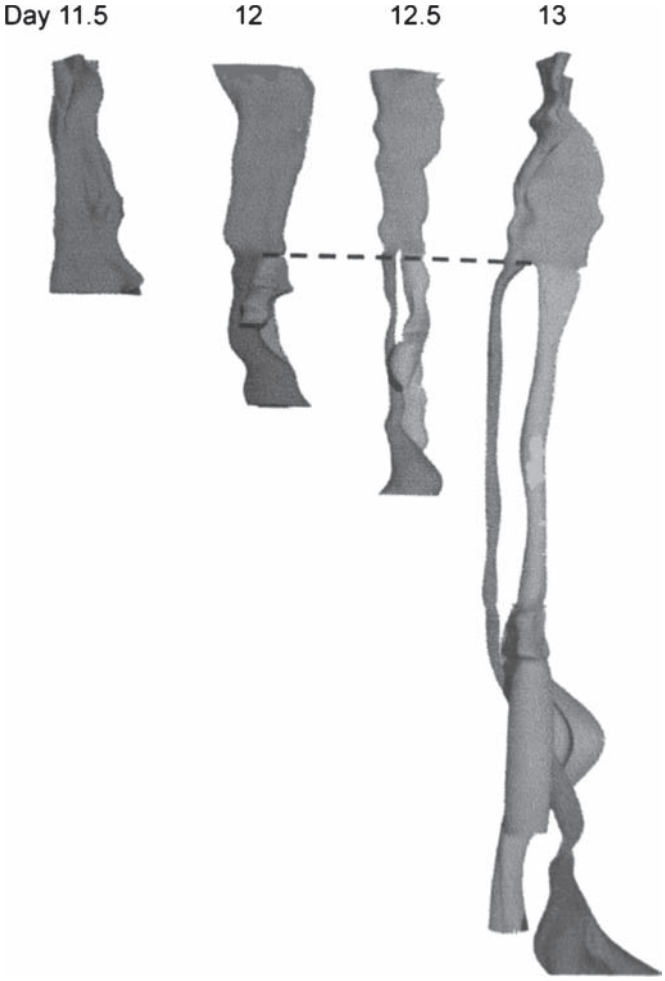


FIG. 22.3. Serial 3-D reconstructive images showing progressive tracheoesophageal separation and esophageal lengthening during normal development of the primitive foregut in the rat (courtesy of Dr. Andrew Williams)

The Upper Esophageal Pouch

In esophageal atresia the proximal esophageal pouch may have an origin different to that of the distal esophagus.[34] For example, in the rat Adriamycin model the proximal esophagus first appears as a dorsal outpouching of the proximal foregut immediately cranial to an area of apoptosis in the dorsal epithelium of the distal pharynx. Subsequently, this elongates through a process of cellular proliferation. Moreover, the proximal esophageal pouch appears to differ significantly in its cellular properties and in its innervation and intrinsic nerve supply.[35,36] In the human the upper esophageal segment may continue to grow and elongate after birth, irrespective of bouginage, perhaps representing a postnatal continuation of the same process.

Role of Sonic Hedgehog and Other Genes

For some years it has been known that the hedgehog signaling pathway plays a crucial role during embryogenesis.[37,38] The sonic hedgehog gene is expressed in the notochord, floor plate, and endodermal epithelial organs, including the trachea, lungs, and digestive tract. Sonic hedgehog is involved in first phase signaling from endoderm to mesoderm.

Sonic hedgehog protein is expressed during foregut development but the levels decrease after the foregut has differentiated into esophagus, trachea, and lungs.[32] Once the organs have formed, the Shh-Gli pathway is turned off permanently. Levels of sonic hedgehog protein in animals exposed to Adriamycin that develop esophageal atresia are markedly lower than in control embryos and do not change during embryonal development[39] (Fig. 22.5). Likewise, in situ hybridization shows that the pattern and levels of sonic hedgehog gene expression are affected by exposure to Adriamycin in rats that develop esophageal atresia.[40] In the human at birth, Shh is expressed in the proximal esophageal pouch but not in the distal fistula.[41]

Mice with a targeted deletion of sonic hedgehog (*Shh* –/– mice) develop lung hypoplasia and various defects of the trachea and esophagus.[38,42] *Gli* mutant mice get esophageal atresia and tracheoesophageal fistula and other abnormalities of the VATER association.[43] The observations indicate a role for *Shh* and *Gli* in endodermal signaling essential for the development of these foregut structures. Three genes, muta-

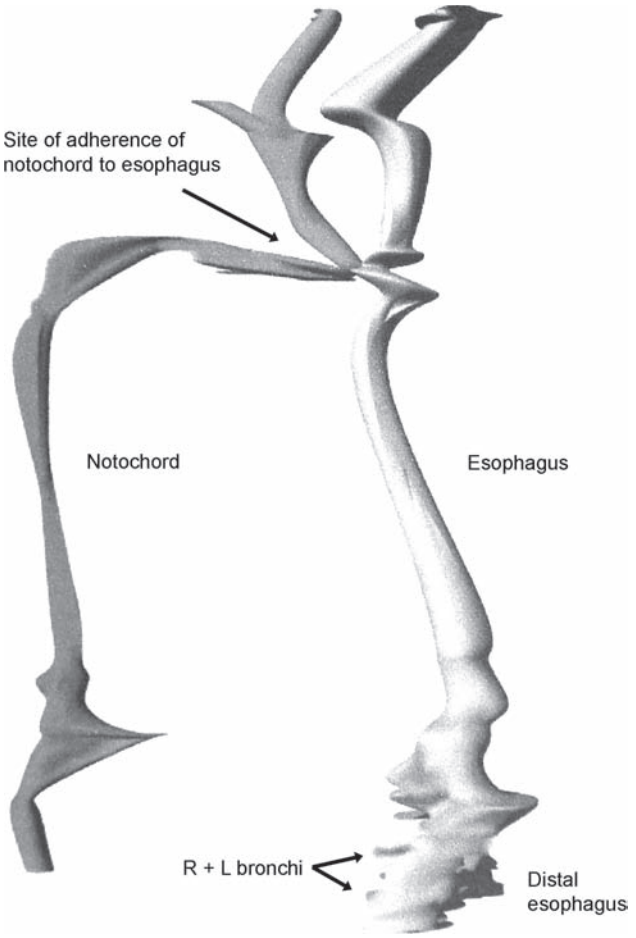


FIG. 22.4. Abnormal notochord in a rat developing esophageal atresia as demonstrated on 3-D reconstructive techniques of serial histological sections (courtesy of Dr. Andrew Williams)

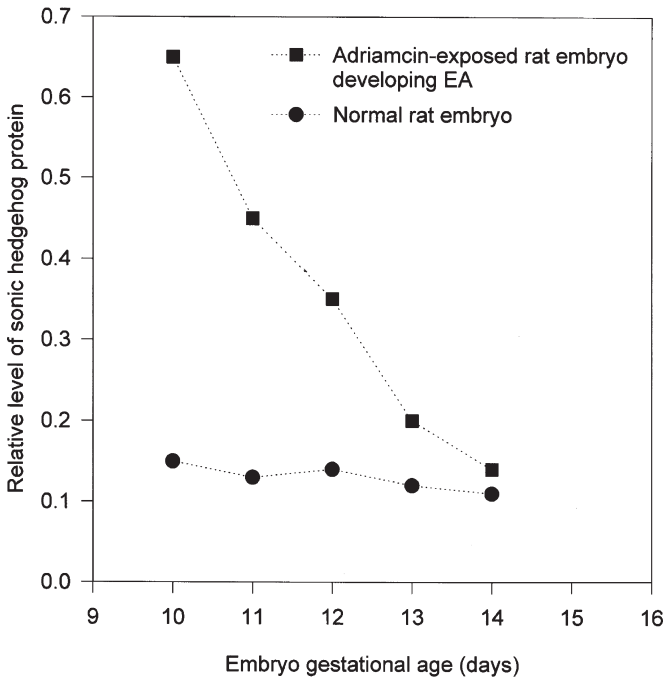


FIG. 22.5. ELISA test reveals that levels of sonic hedgehog protein are significantly lower in rats that are developing esophageal atresia

tions of which cause syndromic forms of esophageal atresia, are transcription factors NMYC and SOX2, and CHD7 that is encoded by a chromodomain helicase DNA-binding gene important for chromatin structure and gene expression.[44]

Anatomical Considerations

Upper Esophageal Segment

Swallowing begins at about 14-weeks gestation and by term several hundred milliliters of amniotic fluid is swallowed each day. The fluid swallowed in esophageal atresia collects in the upper pouch, which becomes dilated and relatively thick-walled. The length of the upper pouch is variable but often extends to within 1 cm of the arch of the azygos vein. The upper pouch can be identified at operation if the anesthetist introduces a stiff catheter into the esophagus.

Lower Esophageal Segment

The distal tracheoesophageal fistula usually commences proximal to the carina of the trachea and, although it may contain tracheal elements near its junction with the trachea,[45] the fistula rapidly becomes esophageal in appearance. It has a caliber significantly smaller than that of the upper esophageal segment. At operation identification of the distal esophagus is assisted by recognition of the vagus nerve coursing over its surface.

Innervation of the Esophagus

The esophagus is innervated by the autonomic nervous system. The sympathetic supply arises from preganglionic neurons in the thoracic and upper lumbar spinal cord with postganglionic fibers entering the esophageal plexuses by visceral branches of the sympathetic trunks and by branches of the greater splanchnic nerves.[46]

The parasympathetic neurons are located in the nuclei of the vagus with the long preganglionic fibers passing within the vagus nerves. They synapse with short postganglionic neurons in the intramural myenteric and submucosal plexuses. These provide innervation to the smooth muscle and secretary cells.

The degree to which esophageal dysfunction is the result of surgical trauma to vagal fibers or to intrinsic abnormalities of the esophagus is debated. The Adriamycin-induced rat model of esophageal atresia has provided some confirmation that the parasympathetic supply is inherently abnormal in esophageal atresia both in terms of the distribution and density of intramural myenteric and submucosal plexuses.[36] Similarly, studies of esophageal function prior to repair of esophageal atresia have shown a positive basal tone and motor incoordination of both proximal and distal esophageal segments, which implies an inherent abnormality of esophageal function.[47] Even patients

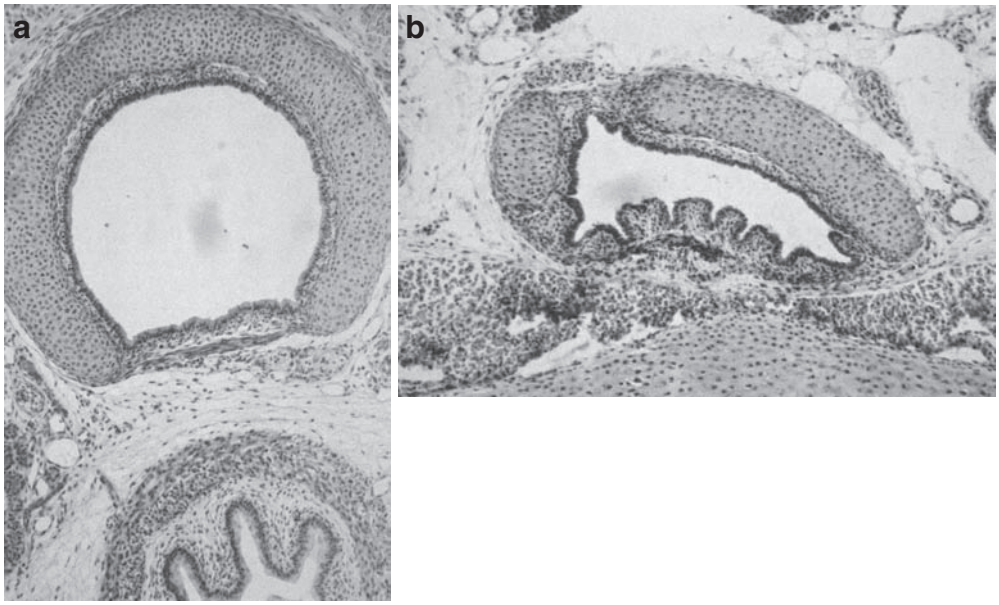


Fig. 22.6. Tracheomalacia in the rat model of esophageal atresia. (**a**) Appearance of the normal trachea in cross section. (**b**) Appearance of the trachea in esophageal atresia. There is disruption of the cartilaginous rings and the posterior membranous part of the trachea is broader, cotributing to collapse of the airway during expiration

with H-type tracheoesophageal fistula may show a degree of esophageal dysmotility.[48,49]

Postoperative manometric studies have demonstrated abnormal peristalsis involving the whole length of the esophagus[50–55] and abnormal lower esophageal sphincter function. In addition, intraesophageal pH studies have shown a high incidence of gastroesophageal reflux and abnormal acid clearance from the esophagus.[56,57] Preoperative cinefluorographic studies have produced similar findings.[48,58,59] Thus, it would seem that there is an inherent abnormality of esophageal function prior to surgery. However, it is likely that inadvertent damage to vagal fibers during dissection of the esophagus will exacerbate the dysmotility.

Blood Supply to the Esophagus

The upper esophageal segment has a good blood supply that predominantly arises from the inferior branches of the inferior thyroid artery, a branch of the thyrocervical trunk. That its branches run vertically downward explains why the upper esophageal segment can be fully mobilized to increase its length without rendering it ischemic.

The blood supply of the distal esophagus is more precarious. It is supplied by segmental esophageal branches from the aorta, which are of relatively small caliber and which anastomose richly with adjacent branches from intercostal and bronchial arteries. Some of these vessels are deficient in esophageal atresia.[60] The distal esophagus is supplied in large part by the ascending branch of the left gastric artery with some support from the inferior phrenic artery. In esophageal atresia this supply is no different from that of the normal esophagus.[61]

The Trachea

The trachea is almost always abnormal in esophageal atresia. Deficiency in the amount of cartilage and an increase in the width of the membranous part produces a structural and functional weakness of the trachea called tracheomalacia.[62] The severity and extent of tracheomalacia in esophageal atresia is variable. Occasionally it may involve the whole trachea, but more often predominantly affects its lower half. The same pathological features of deficiency of cartilage, loss of cartilaginous ring integrity, and expansion of the posterior membranous component are seen in humans and in animal models of esophageal atresia (Fig. 22.6).[63] It is likely that the trachea is inherently abnormal as part of the regional defect. The former belief that it is primarily due to external pressure from the dilated proximal esophageal segment in utero[64] has not been substantiated.[65] Aberrant major vessels may exacerbate the severity of tracheomalacia locally in the adjacent trachea.[65]

Epidemiology

The birth incidence of esophageal atresia and/or tracheoesophageal fistula is between 1:3,448 and 1:4,500 births.[66,67] The prevalence of the abnormality is about 2.8 per 10,000 births.[68]

Twins (or higher multiple births) are more likely to have esophageal atresia.[66,68,69] Chromosomal anomalies have been reported in 6–10% of infants with esophageal atresia,[70,71] although the true incidence is likely to be higher and will become evident as chromosomal analysis becomes more sophisticated. Trisomy 18 and 21 are the most common major chromosomal abnormalities[70] but a wide variety of other chromosomal defects have been identified. Most cases of

esophageal atresia appear to be sporadic, with a recurrence rate of between 0.5 and 2%[66,72] for parents with one affected child, and the empirical risk for an affected child born to an affected parent is 3–4%.[72]

Antenatal Diagnosis

The diagnosis of esophageal atresia is being made with increasing frequency on routine antenatal ultrasonography. The likelihood of esophageal atresia being present is increased when there are maternal polyhydramnios or other abnormalities identified on ultrasonography that are known to be associated with esophageal atresia; these include congenital heart disease, urinary tract abnormalities (e.g., hydronephrosis), and other abnormalities of the VATER association.

Specific ultrasonographic features suggestive of esophageal atresia include a distended upper esophageal pouch, a small stomach, or abnormal swallowing.[73,74] The dilated upper esophageal pouch may vary in volume according to fetal swallowing.[74] A blind upper pouch can be seen on multiplanar ultrasonography from 23-weeks gestation.[73] Demonstration of a patent esophagus may be achieved using a high-resolution linear transducer in fetuses with suspected esophageal atresia.[75] Magnetic resonance imaging has been used to confirm esophageal atresia where ultrasonography is equivocal.[76]

Postnatal Diagnosis

Clinical Features

The classical clinical presentation of an infant with esophageal atresia is of an abnormally "mucousy" infant who is drooling excessive amounts of saliva (Fig. 22.7). There may be a history of maternal polyhydramnios and the infant is often born prematurely (Fig. 22.8).

If the diagnosis is not recognized at birth and feeding is commenced, the child may start choking or gagging, develop respiratory distress, aspirate or even become cyanotic; this should immediately alert the clinician to the correct diagnosis. Clinicians should be aware that some very premature infants with esophageal atresia may not appear to secrete much saliva.

Confirmation of Esophageal Atresia

The diagnosis of esophageal atresia is confirmed when a 10G orogastric tube cannot be passed through the mouth into the stomach. The catheter becomes arrested at about 10 cm from the gums (Fig. 22.9). Although some surgeons routinely obtain a plain X-ray of the chest to show where the tip of the catheter lies this is not essential for the diagnosis.

Fluid aspirated from the catheter does not turn blue litmus paper pink as the upper pouch contains saliva alone. If the esophagus is intact and the catheter enters the stomach aspiration of gastric juice would turn the litmus paper pink.

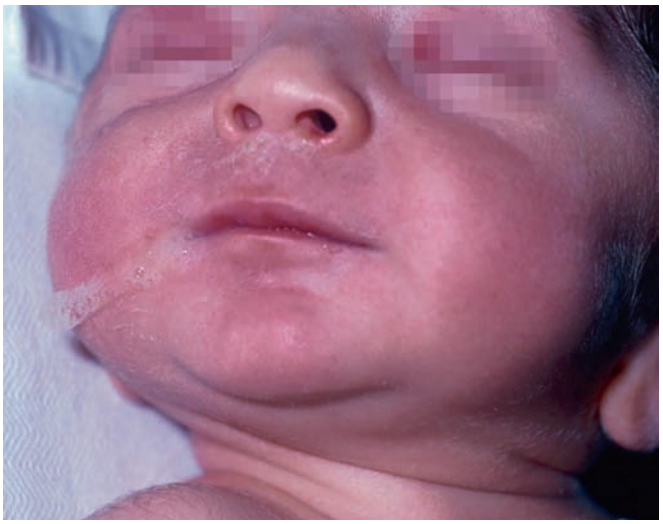


FIG. 22.7. At birth an infant with esophageal atresia typically appears to salivate excessively ("mucousy baby") because saliva accumulates in the blind upper esophageal pouch

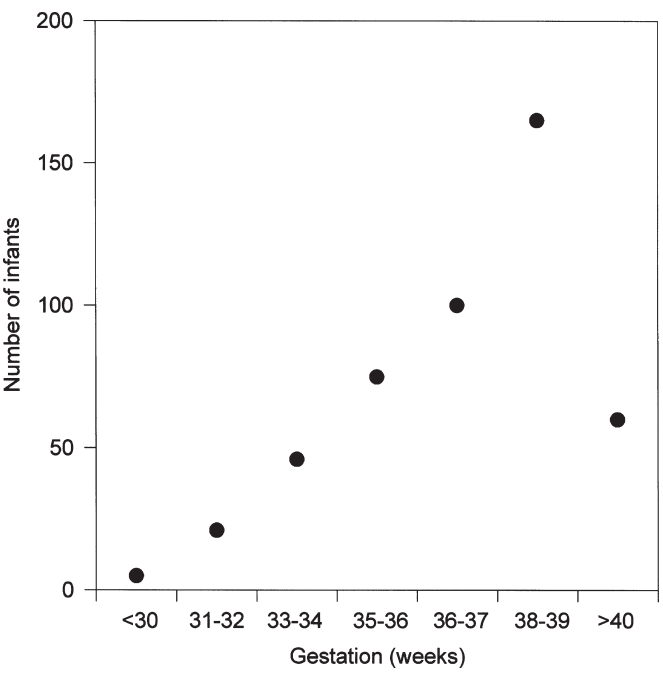


FIG. 22.8. Prematurity is common in infants born with esophageal atresia

A small caliber orogastric tube should not be used because it may curl in the upper pouch giving a misleading impression of esophageal continuity (Fig. 22.10). The catheter is introduced through the mouth rather than the nose to avoid injury to the nasal passages, which are small in the newborn infant. Rarely the oral tube may be passed inadvertently into the trachea and through the fistula into the stomach also giving a false impression of an intact esophagus.[77]

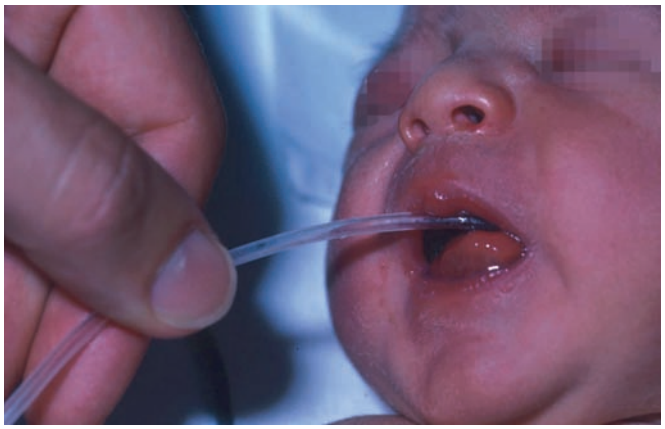


FIG. 22.9. Clinical diagnosis of esophageal atresia is made when a stiff 10G catheter cannot be introduced beyond about 10 cm from the gums

Traumatic introduction of a suction catheter through the mucosa of the posterior wall of the pharynx may give the impression of esophageal atresia when the infant develops copious drooling of saliva and the tube cannot be advanced into the stomach. This is most likely to be seen in premature babies who have required intensive resuscitation.[78,79] The level of obstruction is lower in the chest on a contrast esophagram than would be seen in esophageal atresia.

Routine passage of an orogastric tube into the stomach of all babies at birth is no longer performed because of the potential risks to the baby of traumatically induced apnea by inadvertent injury to the larynx, even though it would enable a diagnosis of esophageal atresia to be made in all babies before a feed is administered. Mediastinal ultrasonography with installation of saliva into the upper pouch,[80] CT scan of the chest,[81] and three-dimensional volume reformatted "transparency" CT images[82] have been used to confirm the diagnosis, but their role, if any, is yet to be established.

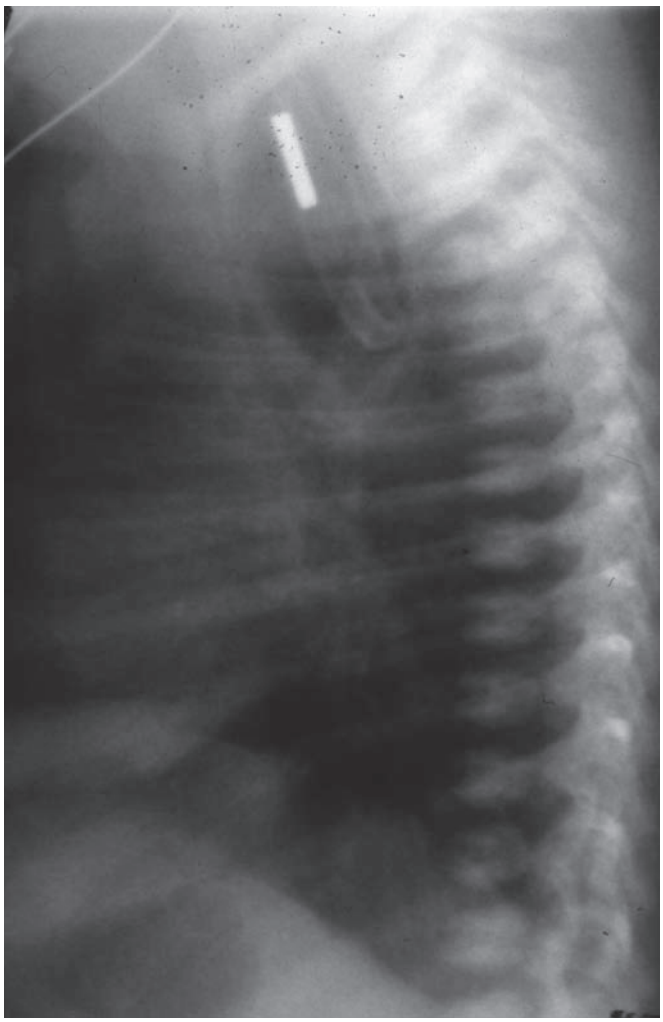


FIG. 22.10. A misleading impression of esophageal continuity can be gained if a small caliber tube is used because it is likely to curl up in the upper esophageal pouch

Determination of the Type of Esophageal Atresia

About 85% of infants with esophageal atresia have a distal tracheoesophageal fistula (Fig. 22.11). The presence of a distal tracheoesophageal fistula can be confirmed by the demonstration of gas in the bowel below the diaphragm (Fig. 22.12). Therefore, the combination of inability to pass a catheter through

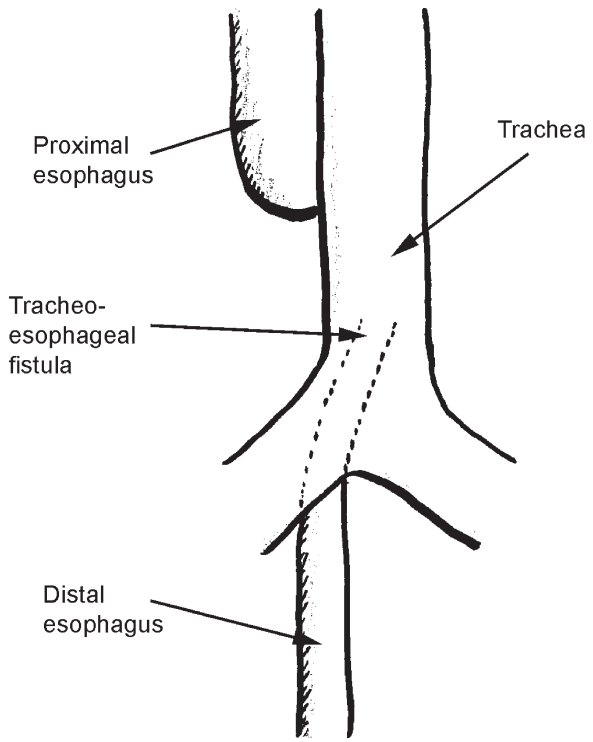


FIG. 22.11. Esophageal atresia with a distal tracheoesophageal fistula. This anatomical variant accounts for approximately 85% of all esophageal atresia and tracheoesophageal fistula cases

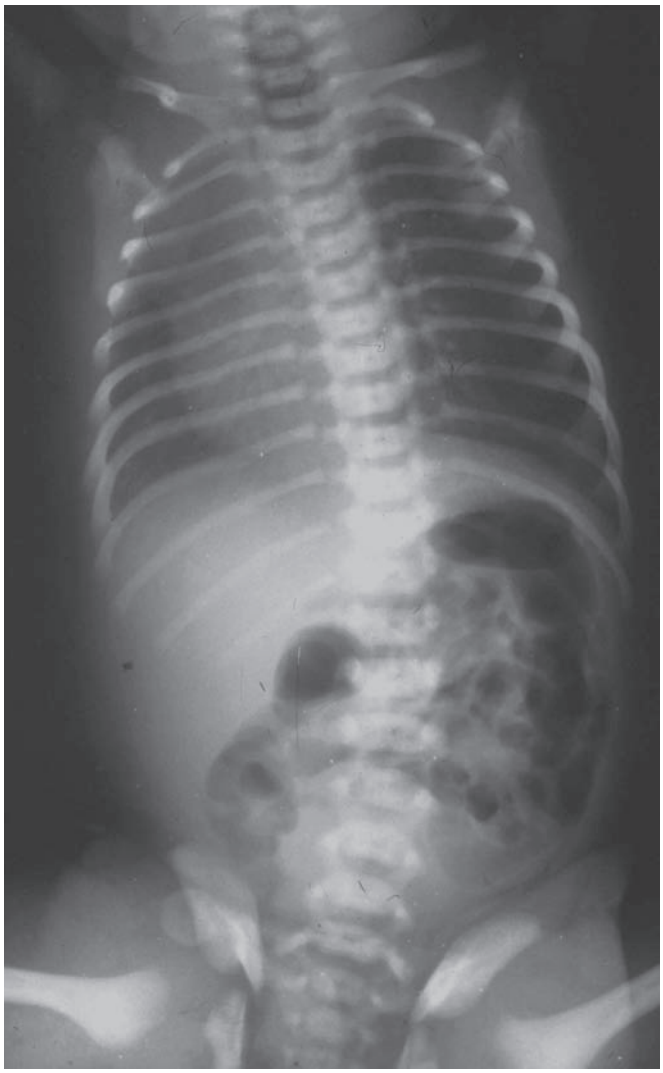


FIG. 22.12. The presence of gas in the bowel below the diaphragam confirms the presence of a distal tracheoesophageal fistula. Air enters the stomach within minutes of birth. Air may also outline the upper esophageal pouch

the mouth into the stomach and the demonstration of gas below the diaphragm is all that is necessary to confirm that the infant has esophageal atresia with a distal tracheoesophageal fistula.

Contrast studies are not required in this situation, although many surgeons perform bronchoscopy routinely prior to repair of the esophageal atresia to do the following:

1. Confirm the level at which the distal tracheoesophageal fistula leaves the trachea (or, rarely, bronchus)
2. Exclude the rare occurrence of a coexisting proximal tracheoesophageal fistula
3. Exclude other abnormalities of the bronchial tree[83–85]

Preoperative tracheobronchoscopy also has been used to cannulate and occlude the distal fistula with a Fogarty catheter to improve ventilation during repair of atresia.[86]

TABLE 22.1. Main abnormalities of the VATER (VACTERL) association.

V	Vertebrae (and rib)
A	Anorectal and other gastrointestinal abnormalities
C	Cardiac
TE	Tracheoesophageal
R	Renal (and urinary tract)
L	Limb, e.g., radial aplasia, thumb abnormalities

Associated Abnormalities

Over 50% of babies with esophageal atresia have other major congenital abnormalities, usually of the VATER (or VACTERL association) (Table 22.1). The CHARGE association (coloboma, choanal atresia, congenital heart disease, and genital anomalies) also occurs with esophageal atresia. A large number of other rare associations have been recognized. Of the many chromosomal abnormalities that have been identified, by far the most common are Trisomy 18 and Trisomy 21.[70] Less common concomitant lesions include anophthalmia,[87–89] diaphragmatic hernia,[90] and cleft lip.

Associated congenital structural abnormalities are the main determinants of survival in esophageal atresia and, of these, congenital heart disease is the most significant.[91,92] Where the prognosis of an associated abnormality (e.g., Trisomy 18, bilateral renal agenesis or hypoplastic left heart) is so poor that long-term survival is impossible, repair of esophageal atresia is not indicated. Consequently, these lesions should be identified before surgery. This is why most centers perform routine preoperative echocardiography and renal ultrasonography. If a major chromosomal abnormality, e.g., Trisomy 18, is suspected clinically urgent karyotyping should be requested. The overall mortality of all patients with esophageal atresia is 10–14%.[92,93]

Congenital Heart Disease

The most common heart defect is a ventricular septal defect and this does not interfere with early repair of the esophageal atresia. In general, correction of the esophageal atresia takes precedence over definitive treatment of the non duct-dependent lesions.[91] Infants with severe right or left obstructive cardiac lesions, in whom either the pulmonary or systemic circulation is duct dependent, may deteriorate rapidly when the ductus closes. Therefore, early identification of duct-dependent congenital heart disease allows a prostaglandin E1 infusion to be commenced prior to repair of the esophageal atresia.[91] Surgery is deferred until the infant is stable. Despite this, the presence of a duct-dependent cardiac lesion is associated with a high incidence of intraoperative critical events and increased morbidity (57%).[94]

Coexistence of Atresias of the Esophagus, Duodenum, and Anorectum

Some children are born with duodenal atresia and an imperforate anus as well. Duodenal atresia can be diagnosed on a

plain abdominal radiograph. The classical appearance of a "double bubble" still occurs because air passes down the distal fistula into the stomach. The anorectal malformation should be evident on inspection of the perineum.

Urinary Tract Abnormalities

The reported incidence of urinary tract abnormalities in esophageal atresia ranges between 10 and 24%.[95–98] Many of the abnormalities require no treatment (e.g., unilateral renal agenesis, duplex kidney) or require treatment but no urgent surgery (e.g., vesicoureteric reflux, pelviureteric junction obstruction). However, bilateral renal agenesis occurs in almost 1% of infants with esophageal atresia. Since this condition is fatal, surgical repair of the esophagus is contraindicated. Patients with bilateral renal agenesis and esophageal atresia often lack the usual features of Potter's syndrome because of the esophageal obstruction. For this reason renal ultrasonography should be performed prior to surgery if the baby has not passed urine.[98]

Management

Antenatal Management

When a diagnosis of esophageal atresia is made on antenatal ultrasonography the parents-to-be should be referred to a pediatric surgeon who can provide accurate information on the nature of the condition and treatment. Where available, the parents-to-be can be introduced to an esophageal atresia support group (vide infra). Knowledge of the diagnosis before birth allows appropriate resuscitation and avoids feeding after birth. The diagnosis is confirmed by attempting passage of an orogastric tube and if the child is born outside a pediatric surgical center early transfer can be arranged. Antenatal knowledge of the diagnosis confers no survival advantage; nor is it of any prognostic significance.[99]

Preoperative Care

The infant is best nursed in an open cot with an overhead heater to allow easy access without excessive heat loss. The upper esophageal segment is kept empty by regular suction. A peripheral line allows intravenous access. Blood is crossmatched but rarely needed. Perioperative prophylactic antibiotics are administered (usually at induction). Vitamin K is given prior to surgery.

The infant is disturbed as little as possible. Excessive crying may increase the infant's oxygen consumption. More importantly, it tends to fill the stomach with air causing abdominal distension with elevation of the diaphragm, which eventually impedes ventilation. The key components of initial resuscitation and management are summarized in Table 22.2. Repair of esophageal atresia is undertaken once resuscitation is complete and the infant is stable.

Repair by Open Thoracotomy

The purpose of the surgery is to divide the tracheoesophageal fistula and to restore esophageal continuity by anastomosing the blind-ending upper esophageal pouch to the proximal end of the lower esophageal segment.

Position

After induction of anesthesia the infant is placed right side uppermost in the full lateral position. A small towel folded beneath the left chest gives lateral flexion and adhesive tape across the right iliac crest secures the patient in position. The right arm is elevated and either attached to a bar just above the head or allowed to lie on the head.

Surgical Approach

A transverse incision is centered just below the angle of the scapula (Fig. 22.13). The fibers of the latissimus dorsi muscle are divided in the line of the incision. The serratus anterior is retracted anteriorly. In the event that posterior fibers of serratus anterior require division to improve access, this is done as low as possible at its origin on the chest wall to avoid its denervation through damage to the long thoracic nerve. The chest is entered through the fourth intercostal space by diathermy dissection of the intercostal muscles. The pleura is gently swept away from the chest wall to provide exposure to the posterior mediastinum. Retraction of the pleura anteriorly exposes the azygos vein. The azygos vein is divided between ligatures (Fig. 22.14).

Division of the Distal Tracheoesophageal Fistula

The fine endothoracic fascia of the posterior mediastinum is incised with diathermy or scissors. The fistula may be seen as it distends with air or by tracing the vagal fibers as they course toward the fistula and continue along the lower esophagus. Care must be taken to avoid damage to these. The angle between the back of the trachea and the fistula is dissected so that the fistula can be transfixed with 4/0 absorbable sutures close to the trachea and divided (Fig. 22.15).

Table 22.2. Key points: initial resuscitation.

Action	Reason
Minimize handling of the baby	Prevent increased oxygen consumption
Keep infant warm	Avoid cold stress
If premature, may need surfactant	Improve ventilation and reduce risk of preferential air entry through tracheoesophageal fistula
Avoid bag ventilation	Prevent excessive air passing through distal tracheoesophageal fistula causing abdominal distension
Regular upper pouch suction	Prevent aspiration of accumulated saliva in upper esophageal pouch
Avoid feeding	Prevent food aspiration
Renal ultrasonography	Confirm functioning renal tissue
Echocardiograph	Exclude duct-dependent congenital heart disease

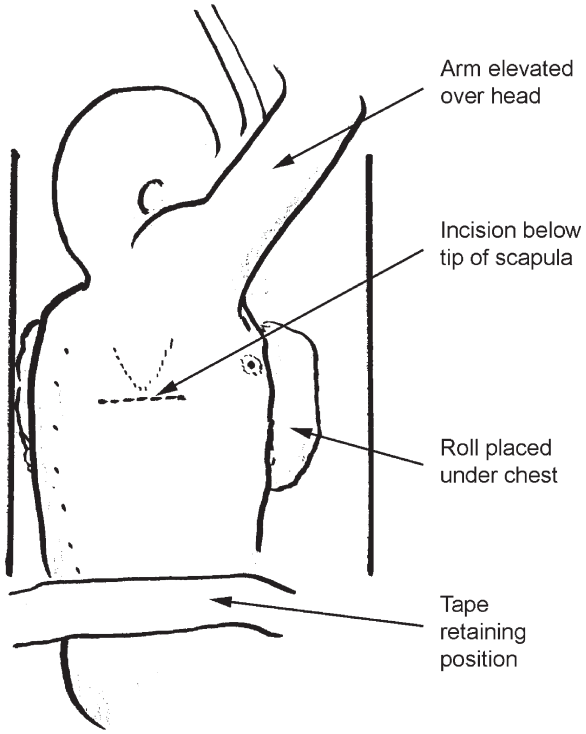


FIG. 22.13. Positioning of the patient for open repair of esophageal atresia. A transverse incision centered just below the angle of the scapula permits an extrapleural approach through the fourth intercostal space

Dissection of the Upper Esophagus

The upper esophageal segment can be identified when the anesthetist pushes downward on a catheter that has been introduced through the mouth into it. A stay suture placed in its most distal part may assist its subsequent mobilization, particularly as it is separated from the trachea. The upper esophageal segment can be fully mobilized well into the neck without concern as to its vascularity.

Anastomosis to Gain Esophageal Continuity

The two esophageal segments are joined by an end-to-end all layered interrupted anastomosis (Fig. 22.16). First, an incision is made through the most dependent part of the blind-ending upper esophagus. Sutures are passed between the esophageal ends, and these must include all layers. Particular attention must be paid to ensure that the mucosa is included, as it tends to retract out of view if there is tension on the esophagus. Once three or four sutures have been inserted in the far wall the esophageal ends are carefully opposed and the sutures secured with the knots tied on the mucosal side. A size 8 French gauge tube is passed across the incomplete anastomosis into the lower esophagus to facilitate completion of the front wall of the anastomosis. A further 5–6 sutures are placed with the knots being tied on the outside. The transanastomotic catheter may be removed when the anastomosis is complete.

Closure

Prior to closure irrigation of the extrapleural cavity with warm saline provides confirmation that there is no air leakage from the trachea. A chest drain is not required unless there is concern about the integrity of the anastomosis. A transanastomotic tube is used only if the infant is very premature and likely to require gavage feeding.

Thoracoscopic Repair

There is increasing experience with the thoracoscopic approach to repair esophageal atresia.[12,13,100–103] The main

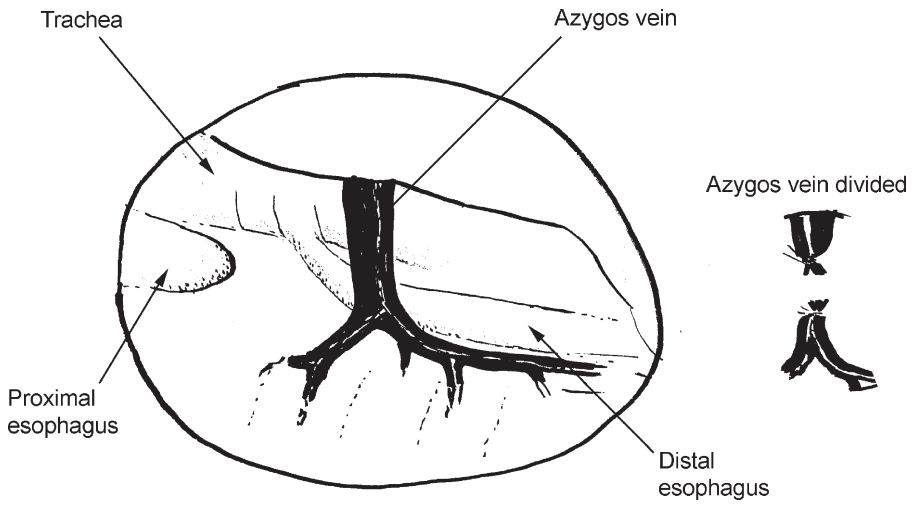


FIG. 22.14. In the extrapleural approach the parietal pleura of the right hemithorax is reflected anteriorly, and the azygos vein divided between ligatures. This exposes the junction between the fistula and trachea

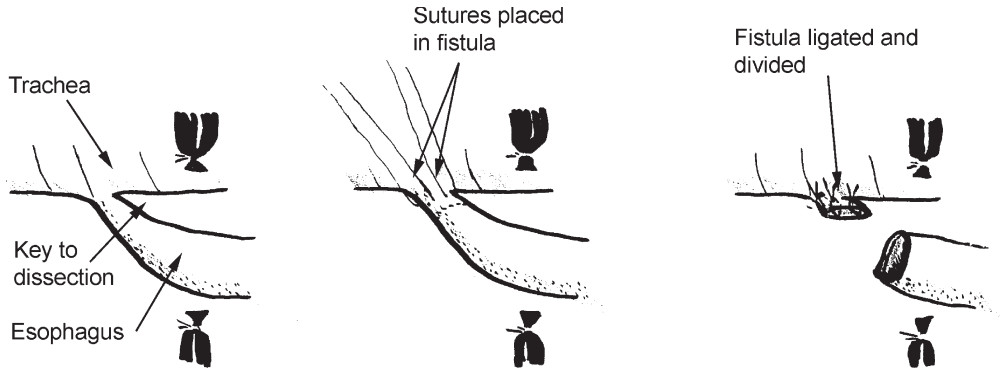


FIG. 22.15. Division of distal tracheoesophageal fistula close to its junction with the trachea

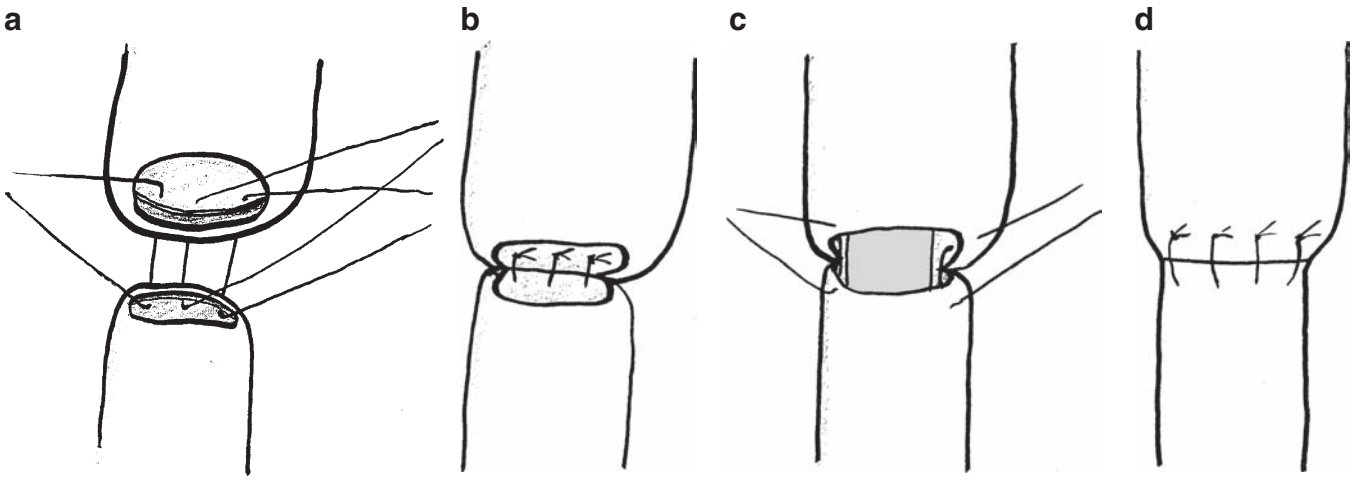


FIG. 22.16. Steps in constructing an end-to-end all layer esophageal anastomosis. (**a**) 3 or 4 sutures are placed in the far wall of both segments. It is important to ensure that mucosa is included in the sutures. (**b**) The posterior wall sutures are approximated simultaneously to draw the two ends of the esophagus together. The sutures are then tied. (**c**) The nasogastric tube is passed across the anastomosis by the anesthetist. Sutures are then placed in the front wall. (**d**) Once the esophageal anastomosis has been completed the nasogastric tube can be removed

advantage of the thoracoscopic approach is magnification of the operative field (Table 22.3). The primary disadvantages are the small working space and the difficulty in performing a delicate anastomosis in a confined area. There may be an increased risk of sutures tearing the esophagus if the anastomosis is under tension because the sutures can only be placed one at a time. Most surgeons use a transpleural approach[13] although an extrapleural approach has also been described.[104]

Not all infants are suitable for thoracoscopic repair. It is contraindicated if the infant is unstable or extremely small. Relative contraindications include congenital heart disease and hyaline membrane disease when the infant is able to tolerate only short periods of single-lung ventilation during ligation of the fistula.[105] Single-lung ventilation can be achieved by selective intubation of the left main stem bronchus by the anesthetist. If this is not possible a pneumothorax can be created by insufflation of carbon dioxide to 4 mmHg pressure with a flow of 1 l/min, which should collapse the lung sufficiently to provide adequate visualization.

TABLE 22.3. Advantages and disadvantages of the thoracoscopic approach to esophageal atresia

Potential advantages of thoracoscopic approach	Potential disadvantages of thoracoscopic approach
Cosmetic appearance (minimal scarring)	Transpleural, should an anastomotic leak occur
Magnification provides excellent visualization	Small operative space
Uniform collapse of lung eliminates retraction trauma	Single-lung ventilation not always achievable in neonates
Less postoperative pain	Thoracoscopic approach requires good lung compliance

Position and Port Placement

The patient is placed in a modified prone position with the right side elevated to approximately 30–45° (Fig. 22.17). Initially a 5-mm port is placed in the fifth intercostal space in the posterior axillary line and a 30° scope is introduced. Two working ports are then placed in the mid-axillary line one or

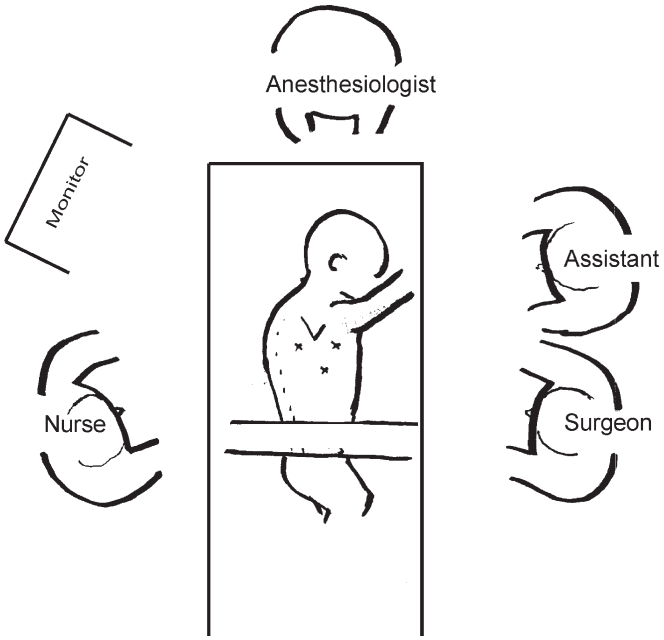


FIG. 22.17. Theatre layout and patient position for thoracoscopic repair of esophageal atresia

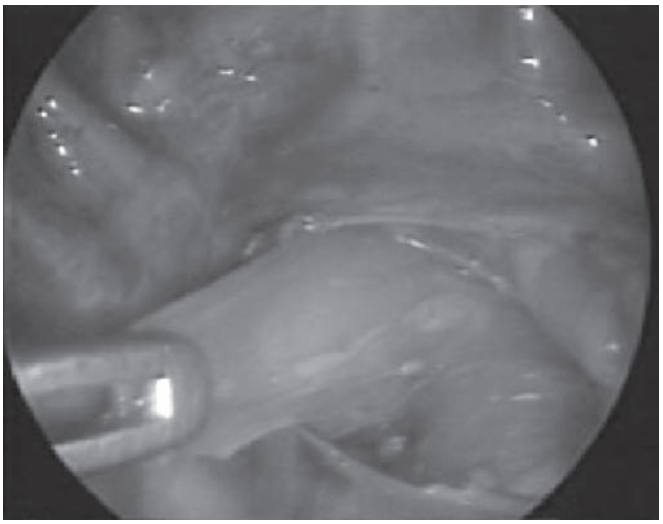


FIG. 22.18. View toward the thoracic inlet during thoracoscopic mobilization of the upper pouch in esophageal atresia (courtesy of Dr. Hossein Allal)

two interspaces above and below the camera port. The upper port is 5 mm to allow for a clip applicator and suture. Sometimes a fourth port is required to help retract the lung.

Closure of the Tracheoesophageal Fistula

The chest is insufflated and the lung collapsed. The azygos vein is divided. The lower esophageal segment is identified and traced in a proximal direction to where it enters the back wall of the trachea. A 5-mm endoclip can be applied on the tracheal side and the fistula is then divided with scissors.

Mobilization of the Upper Pouch

Downward pressure applied on the orogastric tube by the anesthetist helps identify the upper pouch. The pleura overlying the pouch is incised and the pouch mobilized toward the neck with a combination of blunt and sharp dissection (Fig. 22.18). Mobilization can be continued into the thoracic inlet, to gain additional esophageal length, depending on the gap that has to be overcome to achieve an esophageal anastomosis. The most dependent part of the upper esophageal pouch is opened.

Esophageal Anastomosis

A 4/0 or 5/0 absorbable suture is used for the anastomosis. Interrupted all-layer sutures must be placed carefully to ensure mucosa-to-mucosa approximation. Reasonable-sized bites need to be taken to prevent the sutures from tearing out. The knot is most easily tied extracorporally using a knot pusher. Once the far wall of the anastomosis has been completed a catheter introduced through the mouth is advanced into the distal esophagus. This facilitates completion of the anastomosis.

Postoperative Management

Oral feeds (preferably breast milk) are commenced at 2–3 days if the child is stable and well. Some surgeons routinely obtain a contrast study prior to commencing feeds to confirm that there is no anastomotic leakage. However, this may not be necessary where the anastomosis has been constructed without undue tension and looks secure at the time of surgery. Routine dilatation after repair of esophageal atresia is not indicated.[106] Infants with esophageal atresia who have no other major congenital abnormalities can usually be discharged home at about 5 days. Premature infants can be fed through a soft gavage feeding tube passed through the anastomosis, either at the time of surgery or subsequently.

Role of Postoperative Ventilation and Neck Flexion

Postoperative ventilation and paralysis were first advocated for use in low-birth-weight infants likely to develop respiratory difficulties in the immediate postoperative period.[107] Later, its indications were broadened to include those with wide-gap esophageal atresia[108] as this represented those patients where tension on the anastomosis was likely to be the greatest. Subsequently, Davies and Beale[109] and Al-Salem et al.[110] further expanded the indications for nonreversal of anesthesia to include those following a standard uncomplicated repair of esophageal atresia with distal fistula and short gap. The assumption, unproven, was that postoperative ventilation might decrease the anastomotic leakage rate because neck flexion would reduce the tension on the anastomosis and

postoperative ventilation allowed the neck to be kept flexed. The only study of esophageal compliance and the influence of posture on anastomotic tension used a pig model. The authors demonstrated that almost 80% of esophageal lengthening occurs between full flexion and the neutral position, contributing to an increase in esophageal length of about 9%. Compliance of the esophagus appeared to be a linear function of the natural logarithm of the tension applied.[111] Their data implied that any postural change that allowed the esophagus to shorten produced an exponential reduction in the tension at the anastomosis. For this reason the paralysis with cervical flexion may have some application in long-gap esophageal atresia. There is still no evidence that it needs to be performed as a routine.[112]

Surgical Techniques for Long-Gap Esophageal Atresia

A long gap between the esophageal ends is likely when there is no distal tracheoesophageal fistula. The clue to this is absence of gas in the bowel below the diaphragm on plain X-ray of the abdomen (Fig. 22.19) and implies one of two anatomical variants:

1. Esophageal atresia without any tracheoesophageal fistula (75–80%) (Fig. 22.20)
2. Esophageal atresia with a proximal tracheoesophageal fistula (20–25%) (Fig. 22.21)

Some surgeons extend the use of the term "long-gap" esophageal atresia to include esophageal atresia with a distal tracheoesophageal fistula where the upper pouch is relatively short and the fistula arises from the level of the carina or below. Admittedly, in some of these patients there may be several centimeters between the esophageal ends, but usually conventional early primary repair is still quite feasible. True "long-gap esophageal atresia" is a term that is probably best reserved to describe the situation where there is no distal tracheoesophageal fistula, and the blind esophageal ends are almost always widely separated. In the absence of a fistula the distal esophagus tends to be very short, and the distance between the esophageal ends is sufficiently great that immediate esophageal anastomosis is either not possible or extremely difficult. In this chapter the term "long-gap esophageal atresia" is restricted to denote the situation where there is no distal tracheoesophageal fistula and the gap between the esophageal ends is substantial, often precluding early primary definitive repair.

A proximal tracheoesophageal fistula occurs in 20–25% babies. This can be identified either by performing an upper pouch contrast study to demonstrate a communication between the upper esophagus and trachea (which has a risk of aspiration) or by performing flexible bronchoscopy while maintaining spontaneous breathing.[85] Spiral CT with three-dimensional reconstruction may help determine the extent of gap prior to surgery,[113] but is a technique not widely used.

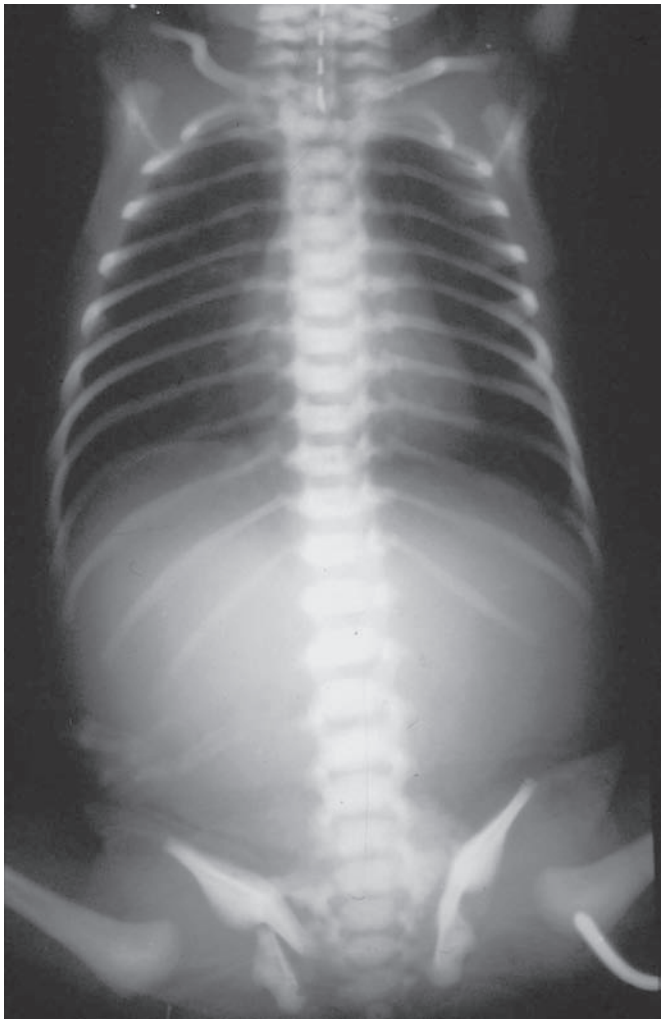


FIG. 22.19. Absence of gas in the bowel below the diaphragm in an infant with esophageal atresia suggests that there is no distal tracheoesophageal fistula

Initial Management of Esophageal Atresia and a Gasless Abdomen

An algorithm for the investigation and management of an infant with an esophageal atresia and gasless abdomen is provided in Fig. 22.22. Until esophageal continuity has been achieved it is important to ensure that the upper pouch is kept free of secretions by regular suction on an ongoing basis: this avoids aspiration if saliva.

Initial investigation is directed at determining whether there is a proximal tracheoesophageal fistula. This is achieved by bronchoscopy or a mid-esophageal contrast study. If there is a proximal tracheoesophageal fistula it should be divided early. Usually esophageal continuity is established at the same operation.

In the more common situation there is no proximal tracheoesophageal fistula. At the time of initial gastrostomy, an

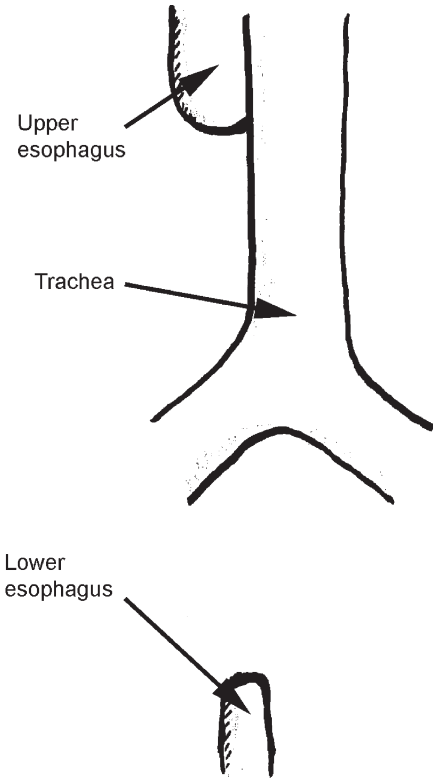


FIG. 22.20. "Isolated" esophageal atresia. There is no tracheoesophageal fistula. In this variant of esophageal atresia the gap between the esophageal ends is usually extensive and the stomach is small

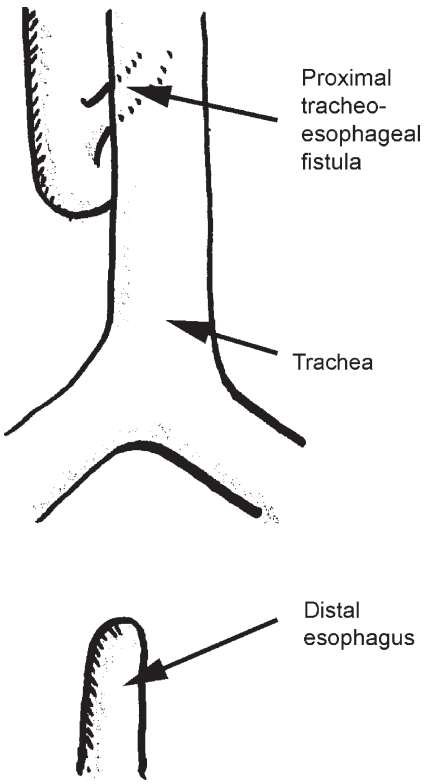


FIG. 22.21. Esophageal atresia with a proximal tracheoesophageal fistula is a less common cause of a gasless abdomen. Again, the gap between the esophageal ends can be extensive

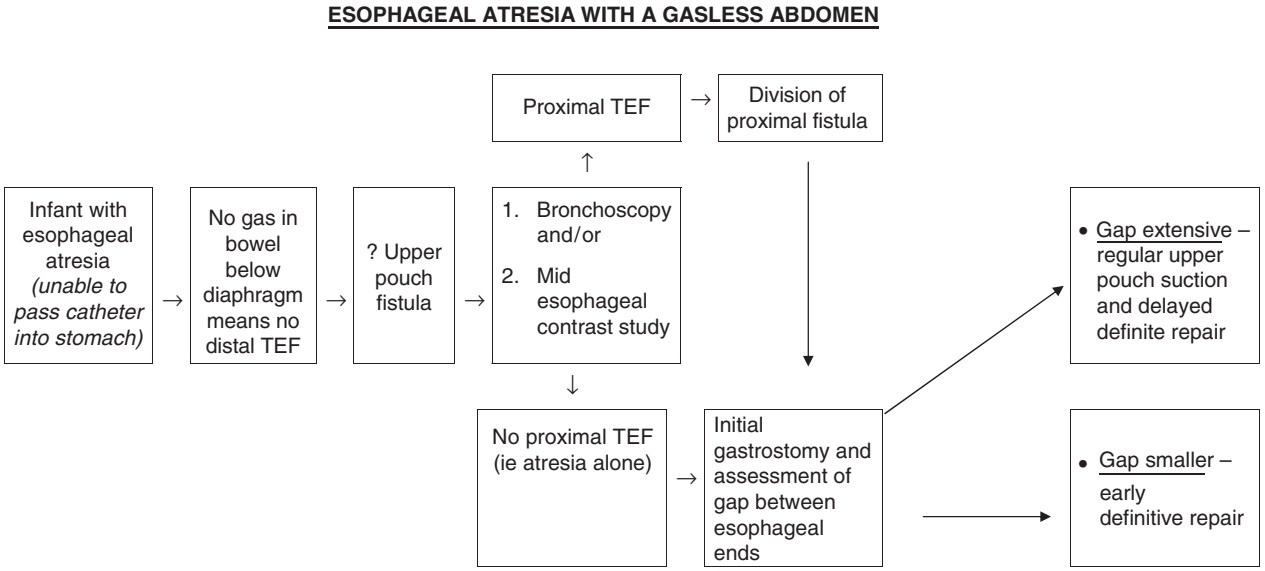


FIG. 22.22. Algorithm for the management of an infant with esophageal atresia and a gasless abdomen

assessment of the distance between the esophageal ends is made. This is done by introducing a metal bougie or sound through the gastrostomy and into the lower esophageal segment, exerting gentle pressure upward at the same time as the anesthetist inserts a radiopaque-tipped flexible catheter through the mouth into the upper esophagus. Fluoroscopic imaging will demonstrate the gap between the two ends and allow a decision to be made as to whether it is safe to proceed to immediate thoracotomy and primary end-to-end anastomosis.[114]

If it is judged that the ends cannot be anastomosed without undue tension because the gap is extensive (often more than four vertebral bodies) regular upper pouch suction is reinstituted and the definitive repair delayed for 1–3 months.

Surgical Techniques Used for Long-Gap Atresia

An extensive gap between the esophageal ends may pose a significant technical challenge. The initial step involves extensive mobilization of the upper esophageal segment into the neck (via the thorax) followed by mobilization of the lower segment down to the diaphragm. These two maneuvers may enable the esophageal ends to be brought together, albeit under considerable tension.

However, there are times when full and extensive mobilization of the esophageal ends remains insufficient to get the esophageal ends together, and other measures to "bridge" the gap are required (Table 22.4). In this difficult situation there are a number of options available, each of which has its protagonists. The relative infrequency of this scenario in any one surgeon's experience, and the absence of multicentered controlled studies, has meant that there is limited information on the relative merits of each. Of the various methods currently employed the following are the most established:

Modified Scharli Technique

This enables extra length of lower esophagus by first dividing the left gastric artery, which allows the lower esophagus to be brought another centimeter or so into the chest, relying on the residual blood supply through the short gastric arteries.[115] The additional maneuver of partial transverse division of the fundus from the lesser curve to create an extension of the lower esophagus, again relying on the short gastric vessels, allows additional length to be obtained (Fig. 22.23). Variations of the technique have been described.[116]

Table 22.4. Maneuvers to achieve esophageal continuity in long-gap esophageal atresia.

Full mobilization of upper esophageal segment into neck
Full mobilization of distal segment down to diaphragm
Circular or spiral esophageal myotomy
Augmentation of esophageal length by:
• Scharli technique (division of left gastric vessels, with or without lesser curve transection
• Gastric transposition
• Great curvature gastric tube (reverse or isoperistaltic)

Gastric Transposition

Gastric transposition via the mediastinal route has the following advantages: (a) good blood supply, (b) adequate length can always be obtained, and (c) the procedure is relatively easy to perform. The operation can be performed as a transhiatal gastric transposition through the posterior mediastinum without a thoracotomy, or a gastric transposition via a thoracoabdominal approach.[117,118] Thoracotomy is performed where there are likely to be extensive esophageal strictures, and in some of those with previous esophageal leakage and failed primary anastomoses where careful dissection is required to remove the residual native esophagus. The open operation has been well described by Spitz[117,119] (Fig. 22.24). A laparoscopic approach has also been devised.[120] Overall, the long-term results are satisfactory, although those who have had previous failed attempts at esophageal reconstruction or replacement tend to have more symptoms.[121]

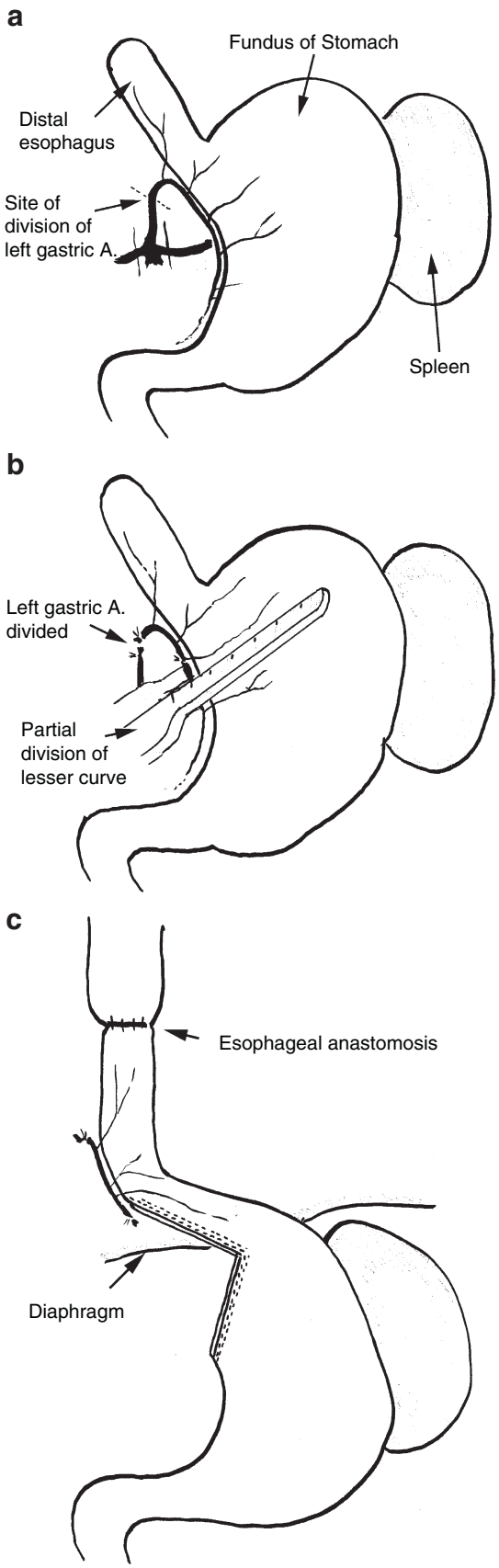
Gastric Tube Procedures

A greater curvature gastric tube of the Heimlich-Gavriliu type[122,123] has stood the test of time and remains a popular method for esophageal replacement. The tube can be based upon the cardia ("reversed gastric tube") or the pylorus ("isoperistaltic gastric tube") (Fig. 22.25). It can be performed safely in infancy.[124] Even though the long-term results of the gastric tube have been better than those of esophagocoloplasty, significant problems may still occur either during the early postoperative period or many years later[125] (Table 22.5).

Other Methods

Over the years a number of other methods have been used with variable success. For example, esophagocoloplasty, a technique that held pride of place for several decades, produced encouraging short-term results, but the relatively high incidence of long-term complications has meant that it is now performed infrequently.

There are several techniques of jejunal transfer[126–129] but none has gained widespread acceptance, perhaps because of difficulties in obtaining adequate length of jejunum without compromising its blood supply, and absence of long-term evidence of advantages over methods using the stomach. Ileal pedicle grafts also have their proponents,[130] particularly for the infant who already has a cervical esophagosotomy and gastrostomy. The technique involves the creation of a Roux-en-Y limb of proximal jejunum advanced through a wide subcutaneous tunnel overlying the sternum. Reconstruction of the esophagus using a free jejunal graft[128,131] has not gained widespread acceptance, and more recently, use of a terminal ileal pedicle graft has been described as well.[130]



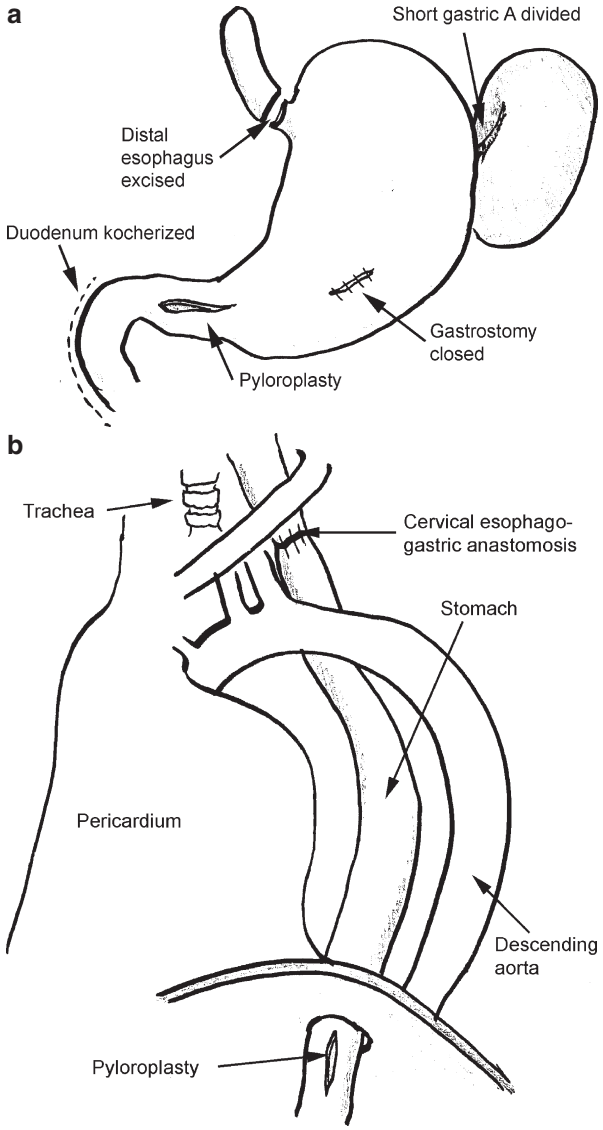


FIG. 22.24. Gastric transposition esophageal replacement. (**a**) Removal of esophageal stump, division of short gastric arteries to mobilize the fundus, closure of the gastrostomy, mobilization of the duodenum and pyloromyotomy. (**b**) The stomach is brought up through the mediastinum to enable an esophagogastric anastomosis to be constructed

FIG. 22.23. Division of the left gastric artery and partial transection of the gastric fundus provides additional length to the lower esophagus. This may permit an end-to-end anastomosis in a child with long-gap esophageal atresia. Scharli also described semifundoplication to restore the angle of His, and pyloromyotomy to improve gastric emptying. (**a**) The anatomy of the left gastric artery. (**b**) Ligation and division of the left gastric artery and partial division of the fundus from the lesser curvature of the stomach. (**c**) The lower esophagus can be mobilized sufficiently to permit anastomosis

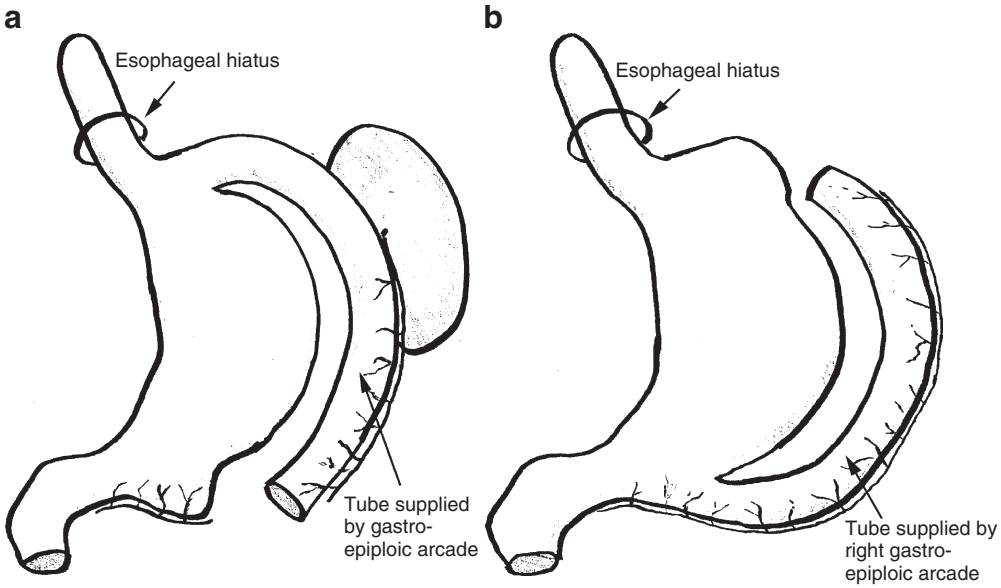


FIG. 22.25. Gastric tube esophageal replacement. (**a**) Gastric tube based on the cardia (reverse gastric tube). (**b**) Gastric tube based on the antrum (isoperistaltic gastric tube)

TABLE 22.5. Postoperative problems following greater curvature gastric tube replacement of the esophagus in esophageal atresia.

Early problems	Late problems
Perioperative mortality	Dysphagia
Necrosis of the graft	Cervical Barrett's
Leakage of the esophagogastric anastomosis	Esophagitis
Dumping syndrome	
Anastomotic stricture (± aspiration pneumonia)	

In the early 1990s circular or spiral myotomies of the upper esophagus became popular. However, this procedure was demonstrated to cause significant injury to the motility and vascular supply of the upper pouch,[132] had a high leakage and stricture rate, and commonly produces a diverticulum[133] (Fig. 22.26). It is now performed rarely.

Another technique described involves raising a flap from the upper segment of the esophagus. The rationale for the anterior mucomuscular flap[134] relied on the observation that the upper esophageal segment had a good longitudinal blood supply and was of greater caliber than that of the lower segment. However, the stricture and leakage rate is significant, and this technique now is only performed in a few centers.

Finally, some surgeons have claimed excellent results by internal or external traction on the esophageal ends, inducing esophageal growth and reducing the gap between the esophageal segments.[135,136] The advantage of these techniques is that eventual esophageal continuity can be achieved without the need for esophageal replacement (with all its shortcomings).

The range of surgical options listed earlier is not an exhaustive testament to the ingenuity of surgeons in devising new ways to overcome this difficult technical challenge. No one current method is clearly superior to the other methods.

Right-Sided Aortic Arch

A right-sided aortic arch may be suspected on plain chest X-ray and confirmed by preoperative echocardiography. In practice, however, the diagnosis of a right aortic arch is often made only at the time of thoracotomy. The arch may obscure the tracheoesophageal fistula and often lies at the level the anastomosis would normally be made. Despite this, where it is encountered unexpectedly, it is still often possible to perform an esophageal anastomosis through the right chest.[137] Sometimes it is better to construct the anastomosis to the right of the right aortic arch. The anastomosis may be technically easier to do in this position, and the aortic arch rarely causes significant compression of the esophagus postoperatively. If it proves too difficult to construct the anastomosis from the right, the infant can be repositioned and a left thoractomy performed under the same anaesthetic.[137] If a right arch is diagnosed with certainty preoperatively, most surgeons repair the atresia through the left chest ab initio.

Coexisting Duodenal Atresia and Imperforate Anus

Where esophageal atresia and duodenal atresia coexist, the esophageal atresia and tracheoesophageal fistula should be dealt with first, following which, a duodenoduodenostomy can be performed under the same anesthetic.[138] Where there is a coexisting anorectal malformation as well, the proximal atresias are dealt with first, followed by a colostomy or definitive

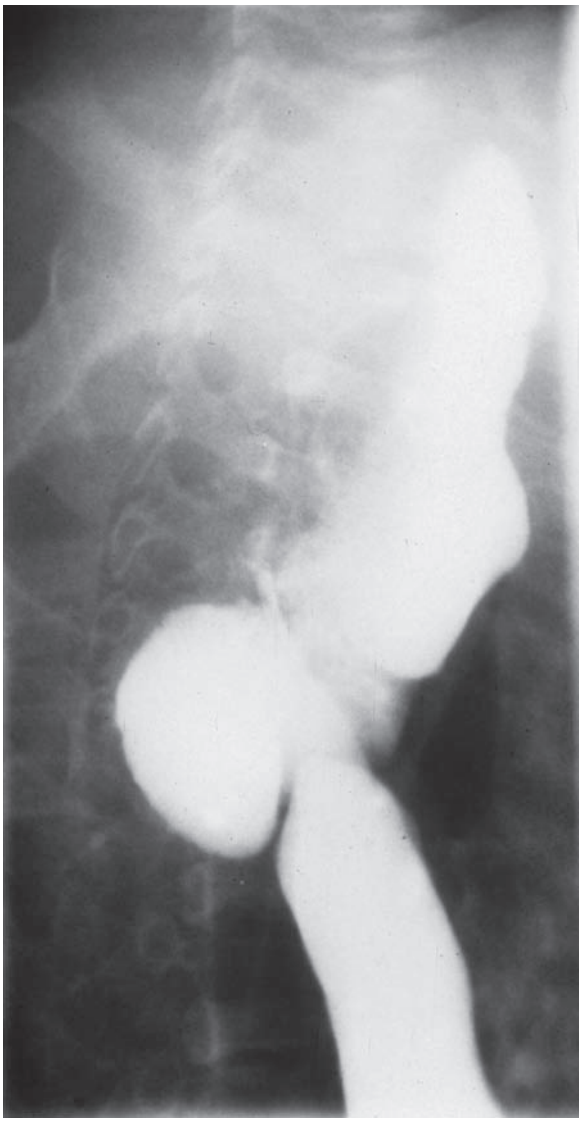


Fig. 22.26. Diverticulum formation following a circular myotomy

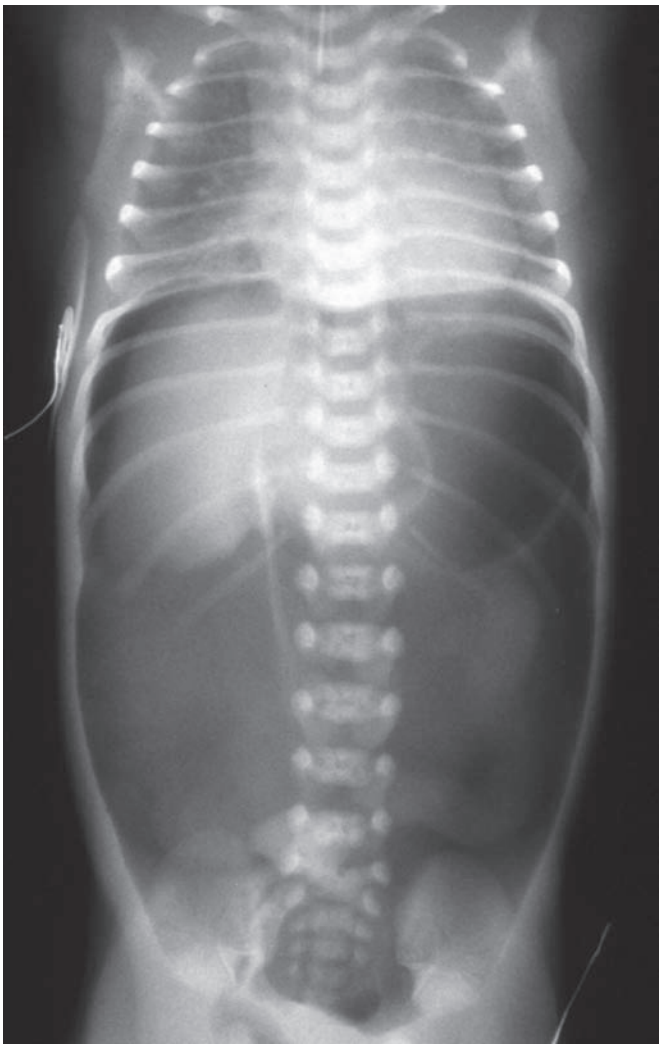


Fig. 22.27. The extremely premature infant with esophageal atresia is at risk of gastric perforation. Gas passes preferentially down the distal tracheoesophageal fistula causing gastric distension. This splints the diaphragm and eventually results in gastric perforation. Note the severe hyaline membrane disease and tension pneumoperitoneum

surgery to the anorectum, depending on the level and severity of the anorectal malformation, the duration of the anesthetic, and the condition of the infant. In any combination of gastrointestinal atresias division of the distal tracheoesophageal fistula is the first priority.

Extreme Prematurity

The extremely premature infant is at risk of developing hyaline membrane disease during the first 24 h after birth. As ventilation becomes more difficult gas tends to pass preferentially through the distal tracheoesophageal fistula. This causes gaseous distension of the abdomen, elevating and splinting the diaphragm and further impeding ventilation. Sometimes, the grossly distended stomach can rupture leading to a tension pneumoperitoneum (Fig. 22.27) and if not dealt with urgently, may progress to hypoxia, cardiac arrest, and death. Surfactant reduces the severity of hyaline membrane disease and the need for ventilatory support, which helps to keep the airway resistance less than that of the fistula.

Where gastric perforation has occurred, immediate insertion of an intravenous cannula into the peritoneal cavity to decompress the pneumoperitoneum should enable the infant to survive until urgent laparotomy is performed. At laparotomy a Foley catheter can be introduced through the gastric perforation, which, conveniently, is usually on the anterior wall of the stomach. The Foley catheter is then directed into the lower esophagus, which it will obstruct, even without inflation

of the balloon. The stability this now achieves allows the tracheoesophageal fistula to be ligated via a thoracotomy.[139] If the infant is sufficiently stable, the esophageal ends can be approximated under the same anesthetic, i.e., complete primary repair is performed. The abdomen is then closed, with or without gastrostomy.

Obviously, there is an advantage to the premature infant if the tracheoesophageal fistula is controlled early. Consequently, division of the fistula (and repair of the atresia) in the premature infant is best performed as soon as the infant is stable and before hyaline membrane disease becomes established.

Tracheomalacia

Infants with severe tracheomalacia may become symptomatic within days or weeks of birth, but generally develop significant symptoms at 2–3 months of age. They have a characteristic harsh barking cough (sometimes called the "TOF cough" or "seal bark cough"). They may be reluctant to feed because of difficulty in breathing during feeds, or have a respiratory arrest or "near miss" sudden infant death. They develop wheeze and intercurrent respiratory infections readily. Most will demonstrate signs of intermittent expiratory obstruction with normal inspiration.[140] All infants with esophageal atresia have some degree of tracheomalacia, although in most it causes few problems and tends to improve with growth.

Indications for Surgery

The child with recurrent cyanotic episodes due to expiratory obstruction who has associated gastroesophageal reflux presents a difficult problem. The extent to which the respiratory obstruction that occurs as a result of the tracheomalacia may induce or aggravate associated gastroesophageal reflux or whether the gastroesophageal reflux is exacerbating the effects of tracheomalacia may be difficult to establish. Some centers prefer to correct the gastroesophageal reflux first in the expectation that the symptoms from tracheomalacia will decrease, whereas other groups proceed directly to aortopexy. Aortopexy is indicated if repeat life-threatening episodes occur.

Aortopexy

Aortopexy involves suture fixation of the aorta to the posterior surface of the sternum, thus elevating the anterior wall of the trachea. It is usually performed through a left thoracotomy and usually provides immediate and permanent relief of obstructive symptoms.[141,142]

The fascia covering the ascending arch of the aorta is opened to expose its adventitial surface, and the reflection of the pericardium onto the ascending aorta is deliberately opened. The exposed aorta can be sutured directly to the posterior surface of the sternum using interrupted sutures or by suturing a Dacron patch to the anterior surface of the aorta and then suturing the patch to the sternum; this draws the ascending arch of the aorta well forward.[143]

Complications

Anastomotic Leak

Leakage from the esophageal anastomosis historically has been a major cause of morbidity after surgery, and has led to several changes in surgical technique – these include the type of anastomosis employed and the extrapleural approach. Leakage may vary enormously in significance, from a minor asymptomatic leak in a well infant only detected on contrast radiology, to complete anastomotic disruption with mediastinitis, empyema, pneumothorax, and septicemia. Assessment of anastomotic leakage must take into account its severity and sequelae (Table 22.6).

A number of factors may predispose to leakage from an esophageal anastomosis (Table 22.7), and recognition of these factors has undoubtedly led to the low leakage rates seen in contemporary practice.[144] The advent of total parenteral nutrition and better antibiotics has allowed a more conservative approach to the management of anastomotic leakage. Even major anastomotic dehiscence can be managed nonoperatively with chest tube drainage, cessation of oral feeds, total parenteral nutrition, and antibiotics.[145] Almost always the leak will close spontaneously. Radical intervention such as cervical esophagostomy is virtually never required and should be reserved for the patient in whom supportive therapy (e.g., total parenteral nutrition) is not available. A longstanding leak may require gastrostomy to allow continuation of enteral feeds. If there is coexisting gastroesophageal reflux, use of a transpyloric tube should be considered so that reflux from the stomach does not perpetuate the leak.

Table 22.6. Management of anastomotic leakage after repair of esophageal atresia.

Incidental finding on postoperative contrast study, no clinical symptoms	Observe No specific treatment Continue oral feeds
Minor leakage: saliva in chest drain (where used), but infant well	Cease oral feeds Antibiotics Will close spontaneously
Major leak: mediastinitis, abscess, pneumothorax, or empyema	Major disruption of anastomosis on imaging Cease oral feeds Antibiotics May require further surgery or drainage Commence TPN

Table 22.7. Factors that predipose to leakage from the esopahgeal anastomosis.

Operative technique	Incorrectly placed sutures
	Insecure sutures
	Sutures "cut through" esophagus
Ischemia	Excessive oesophageal mobilization
	Extreme tension on esophagus
	Esophageal myotomy
Tension	Esophageal ends pull apart
	Gap between ends >4 cm
Infection	
Suture material	Silk

A pseudodiverticulum can occur following leakage from the anastomosis but usually heals satisfactorily without long-term mechanical complications. This should be distinguished from ballooning at the site of a circular myotomy, which also produces a diverticulum.[133]

Esophageal Stricture

An anastomotic stricture is the most common reason for further surgery to the esophagus after repair of esophageal atresia.[144] The factors that influence stricture formation are similar to those that produce anastomotic leakage, with the addition of gastroesophageal reflux as a potent cause of stricture formation. The combination of gastro-esophageal reflux and delayed esophageal clearance (because of esophageal dysmotility) means that the acid may bathe the vulnerable region of the anastomosis for prolonged periods, increasing the likelihood of an anastomotic stricture.

Patients with a stricture develop feeding difficulties and dysphagia. As babies they may appear to be "slow feeders" and have excessive regurgitation, with or without cyanotic spells. Older children may present with foreign body impaction of food in the esophagus, particularly in the 2–5-year age group. The diagnosis can be confirmed by either endoscopy or Barium swallow. Endoscopy is used as the first investigation when the child presents with foreign body impaction.

Radial balloon dilatation under fluoroscopic control is probably the most effective and safest technique of dilatation of an anastomotic stricture.[146] One or two dilatations may be all that is required to treat patients with mild narrowing of the esophagus.

Patients with associated gastroesophageal reflux should be placed on an H_2-receptor antagonist or proton pump inhibitor. If these fail or are not tolerated, consideration should be given to performing a fundoplication at which time a further dilatation of the stricture may be required.

Recurrent Tracheoesophageal Fistula

A recurrent tracheoesophageal fistula remains a severe and potentially dangerous complication of esophageal atresia. Its incidence has now declined to under 2%. It is believed that many recurrent fistulae are caused by an anastomotic leakage that results in infection in the area of the repair, particularly when the site of tracheal closure is very close to the anastomosis.

Recurrent fistulae may appear in the early postoperative period, or several years after surgery. They can present with a range of symptoms including coughing, gagging, choking, cyanosis, apnea, dying spellings, and recurrent chest infections. Usually, however, the child simply coughs and splutters with each feed.

The diagnosis is confirmed on bronchoscopy or cineradiographic tube esophagography with the patient prone (Fig. 22.28). The recurrent fistula usually arises from the pouch of the original fistula.

Spontaneous closure is unlikely to occur. The fistula should be divided when the child is in optimal respiratory and general condition, and this may necessitate a period of total parenteral nutrition. Gastrostomy is now rarely indicated. The conventional approach is through the original right fourth interspace using a transpleural approach. The passage of a fine ureteric catheter through the fistula immediately prior to the thoracotomy may facilitate its localization during the operative repair.

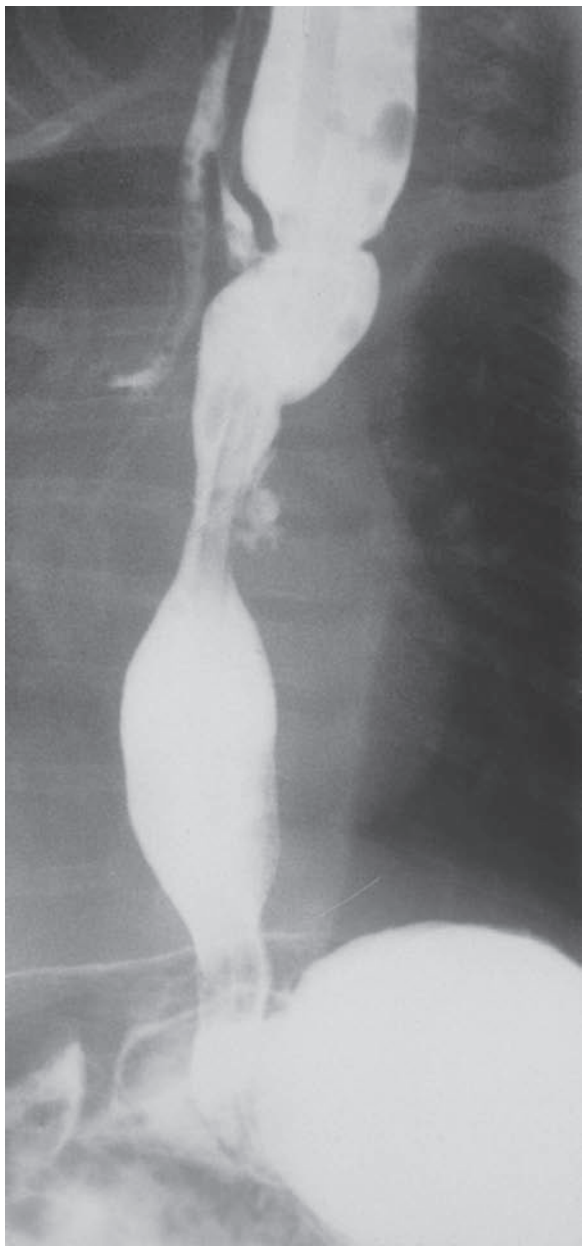


FIG. 22.28. Recurrent tracheoesophageal fistula

There are a number of reports of endoscopic obliteration of recurrent tracheoesophageal fistulae using diathermy obliteration,[147] tissue adhesive,[146] and a combination of a tissue adhesive and sclerosing agent[148] of a coated self-expanding plastic stent.[149] It has been suggested that these techniques may have special application in regions where additional surgery has an unacceptably high morbidity or where parents are reluctant to allow their child to undergo further surgery.[148]

Gastroesophageal Reflux and Esophageal Dysmotility

Gastroesophageal reflux can be troublesome for two reasons:

1. It may lead to aspiration of gastric contents
2. Prolonged exposure of the esophageal mucosa to acid may lead to an anastomotic stricture and Barrett's esophagus

There is a tendency for gastroesophageal reflux to improve with age, but despite this it must be taken more seriously than in the otherwise normal infant. The availability of effective H_2-receptor antagonists and proton pump inhibitors has dramatically reduced the need for early fundoplication. Antireflux surgery is now reserved for those with ongoing episodes of aspiration or recurrent chest infection, or for those in whom medication is not tolerated or has failed.

Laparoscopic antireflux surgery in esophageal atresia has largely replaced open fundoplication.[150] Postoperative pH monitoring identifies most, but not all, infants who will go on to develop esophagitis or require an antireflux procedure.[151]

About 30% of children have short-term dysphagia following fundoplication probably because the increased resistance at the gastroesophageal junction unmasks the preexisting esophageal dysmotility (poor peristalsis and delayed esophageal clearance) that is usual in esophageal atresia. The long-term effects of gastroesophageal reflux into an abnormal esophagus that has poor motility and delayed clearance are discussed in more detail later.

Rare and Unusual Variants

The Congenital "H" Fistula

The isolated tracheoesophageal fistula ("H" fistula) without atresia accounts for about 4% of congenital esophageal anomalies (Fig. 22.29). It presents with entirely different symptoms from esophageal atresia because the esophagus is patent. It is included in discussion of esophageal atresia because of its presumed common etiology.

Figure 22.29 shows the oblique passage of the fistula that runs from the trachea in a caudal direction to the esophagus. The symptoms it produces relate to abnormal passage of air through the fistula from the trachea to the esophagus (and stomach) and of esophageal contents, which may include saliva, gastric juice, and milk, into the trachea.

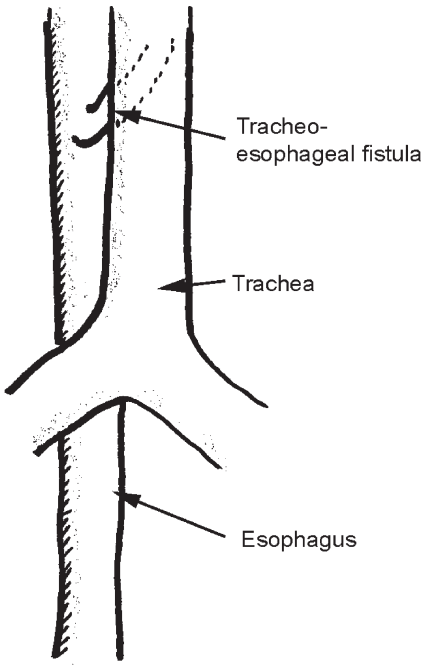


FIG. 22.29. Congenital isolated tracheoesophageal fistula ("H" fistula) without esophageal atresia

Diagnosis

It usually presents in the first few days of life with choking on feeds and unexplained cyanotic spells.[152] Choking attacks may be associated with abdominal distension.

Older infants may present with recurrent bouts of pneumonia, usually involving the right upper lobe, and unexplained intermittent bouts of abdominal distension. Less common features include a hoarse cry and failure to thrive. Some children are months or years old before the diagnosis is recognized.

Investigation

An isolated tracheoesophageal fistula can be diagnosed radiologically or by endoscopy. Video-esophagography (Fig. 22.30 involves introduction of a tube into the mid-esophagus, with the infant lying prone, with injection of contrast as the tube is slowly withdrawn.[153] Familiarity with the technique is important, as a standard barium swallow will miss an H-fistula in 25% of occasions.[154]

The fistula can be seen readily on bronchoscopy as an abnormal opening on the posterior wall of the trachea. Sometimes bronchoscopy is performed immediately prior to the surgery to divide the fistula so that a catheter can be introduced through the fistula to aid in its identification at open surgical exploration.[155]

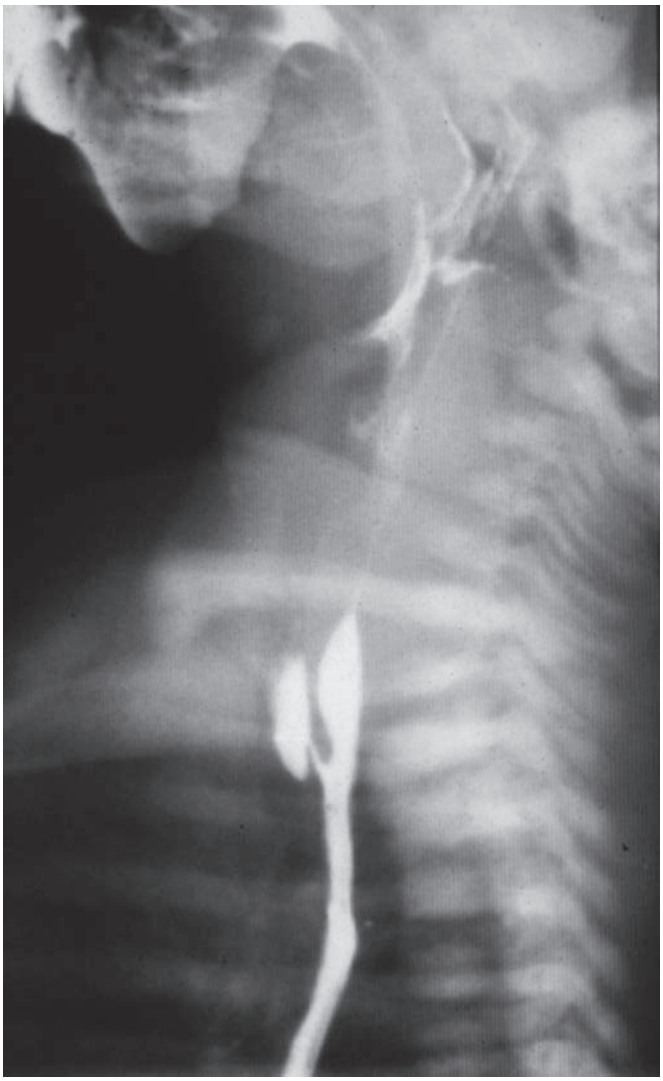


FIG. 22.30. A congenital "H" fistula can be demonstrated by videoesophagography with the infant prone

Operative Management

Almost all H-fistulae can be approached through a right supraclavicular incision.[154,156] The sternomastoid muscle is retracted posteriorly, although division of its sternal head may improve the exposure. The incision is deepened antero-medial to the carotid sheath. The trachea is recognized by palpating its rings. The fistula is found in the groove between the trachea and esophagus, and on its external surface it looks surprisingly short and broad. It is not necessary to dissect completely around the esophagus or to control it with a vessel loop as this increases the risk of damage to the recurrent laryngeal nerve. However, a sling placed around the fistula itself may help its dissection and division. The fistula should be transfixed and divided rather than ligated alone, to reduce the likelihood of recurrence of the fistula. Placement of muscular flaps between the divided ends of the fistula may also decrease the recurrence rate but is probably not required as a routine. Drainage of the wound is not necessary and gastrostomy is not used. The anesthetist should always inspect the vocal cords to confirm their movement at the completion of the operation. Oral feeds are commenced the next day.

Complications

The main complications of the procedure are recurrent laryngeal nerve palsy, either unilateral or bilateral and recurrence of the fistula.[154] Leakage at the site of closure is rare, but may result in mediastinitis, or a recurrent fistula. Other complications are rare and include injury to the esophagus producing mediastinitis or an esophagocutaneous fistula. Pneumothorax and tracheal obstruction can be avoided with good surgical technique. Postoperative aspiration and pneumonia have also been reported.

Other Rare Variants

A large number of rare variants of esophageal atresia have been reported.[157,158] It is not necessary to list (or learn) them all; rather, the surgeon should be aware that bizarre variants do occur, and may be encountered unexpectedly at the time of surgery. The exact anatomy of the variant should be defined, and the abnormality corrected following normal surgical principles.

Congenital Lower Esophageal Stenosis

An encircling "cartilaginous" rest within the wall of the lower esophagus is a rare but well-recognized condition that occurs in 1:25–50,000 births.[159,160] It may produce obstructive symptoms and often occurs in association with esophageal atresia. Between 5 and 14% of infants born with esophageal atresia will also have a congenital stenosis of the esophagus.[161] Less common variants of this lesion also occur (Table 22.8), but all are believed to have a common etiology.

The lesion usually becomes symptomatic at the time of introduction of solid foods, or suddenly when foreign body impaction of the esophagus occurs.

TABLE 22.8. Classification of congenital stenosis of the lower esophagus.

Encircling cartilaginous "rest", i.e., tracheobronchial remnant in esophageal wall (most common)
Fibromuscular thickening (idiopathic muscular hypertrophy)
Membranous web or diaphragm (extremely rare)

Contrast radiology demonstrates an abrupt and fixed narrowing, which, in esophageal atresia, may be difficult to distinguish from a low esophageal stricture secondary to esophagitis and gastroesophageal reflux. High-frequency catheter probe endoscopic ultrasonography can demonstrate the hyperechoic cartilage at the site of esophageal narrowing.[162] The narrowing persists despite attempts at radial balloon dilatation, and definitive treatment involves resection of the affected esophageal segment and end-to-end esophagoesophagostomy.

Prognosis and Long-Term Outcomes

Predictors of Outcome

Over the last 60 years there has been a dramatic improvement in the survival rate of repaired esophageal atresia (Fig. 22.31). Nowadays, all patients with esophageal atresia are expected to survive almost irrespective of their gestation, provided there are no major concomitant congenital malformations.[163] There has been a steady decline in overall mortality due to esophageal atresia throughout the world until about 1985.[163–166] In the early years much of the mortality was the result of respiratory failure, inadequate resuscitation, pneumonia, hyaline membrane disease, and other complications of prematurity. Another major cause of mortality was from complications of the surgery itself, particularly those related to anastomotic dehiscence and poor nutrition. Recognition of this led to the Waterson classification,[167] which used to be valuable in identifying risk factors and in predicting outcome in infants with esophageal atresia. However, it is no longer relevant because mortality is now largely determined by the type of severity of concomitant congenital abnormalities alone.[163]

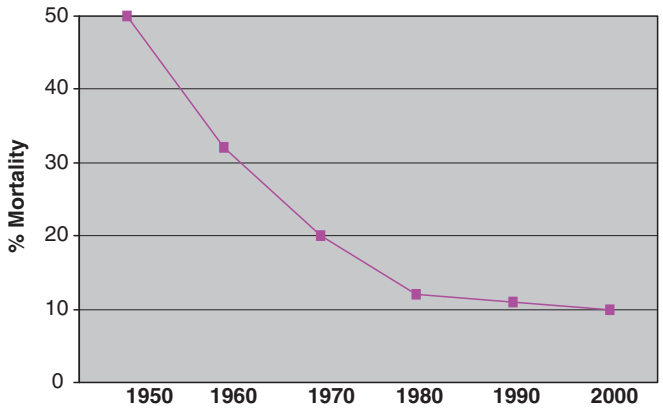


FIG. 22.31. Survival in esophageal atresia, showing the steady improvement in outcome with the passing of each decade. The residual mortality of about 10% is due to congenital abnormalities such as hypoplastic left heart, bilateral renal agenesis, and Trisomy 18

Definitive repair of an esophageal atresia may not be appropriate in patients with an identifiable concomitant congenital abnormality that is known to be lethal or associated with an extremely poor prognosis.

Esophageal Function

The most common long-term problem is dysphagia. It occurs in almost half of all patients.[168] Likewise, gastroesophageal reflux occurs in about 48%.[168] It is likely that all patients have a degree of esophageal dysmotility and poor peristalsis, but it appears to improve with age, or alternatively, patients' eating habits are modified to better accommodate it.

Foreign body impaction is most likely to occur under the age of 5 years, after which time it becomes relatively infrequent. However, most children and adults with esophageal atresia will have their meals with a glass of water and deliberately chew their food well.

Growth

Early studies suggested that growth might be impaired in survivors of esophageal atresia.[169] Other studies have confirmed that some children exhibit early physical developmental retardation but that height and weight eventually become normal.[168,170] By adulthood both height and weight centiles after repair of esophageal atresia follow a normal distribution.[171]

Risk of Esophageal Malignancy After Repair of Esophageal Atresia

Now that some of the older survivors of esophageal atresia are reaching their sixth decade some of the potential long-term sequelae of the condition are becoming evident. There have been three reports of esophageal adenocarcinoma following repair of esophageal atresia.[172–174] Two of these patients were in their 20's and two had areas of Barrett's epithelium identified. It is accepted that gastroesophageal reflux and esophagitis are more common in patients after repair of esophageal atresia than in the general population,[175–177] with a concomitant increase in the incidence of Barrett's esophagus, a known precursor of adenocarcinoma.[177] Esophageal atresia patients appear to be particularly vulnerable to the adverse effects of gastroesophageal reflux because of their esophageal dysmotility and poor esophageal clearance, which means that the time the esophageal mucosa is exposed to acid from reflux is prolonged. Squamous cell carcinoma after repair of esophageal atresia has also been reported.[178] There are two other situations in which malignancy may occur in esophageal atresia patients (Table 22.9). During esophageal replacement surgery using

Table 22.9. Factors contributing to esophageal malignancy after repair of esophageal atresia.

Combination of gastroesophageal reflux and esophageal dysmotility (poor esophageal clearance of reflux acid) leading to Barrett's epithelium
Retained esophageal segment after oesophageal replacement
Squamous cell carcinoma in skin tube conduits

Table 22.10. Esophageal atresia support groups.

Name of group	Country	Contact
International Esophageal Atresia Team		www.tefuater.org/ieat.html
Esophageal Atresia Research Axillary (OARA)	Australia	Holsom@ozemail.com.au
Speiserohrenmi Bbildungen (KEKS)	Germany	Info@keks.org or www.keks.org
Tracheoesophageal Fistula Support Newsletter (CHEW)	UK	Info@tofs.org.uk or www. tofs.org.uk
The VATER Connection	USA	www.vaterconnection.org
EA/TEF Family Support	USA	www.eatef.org
KEKS	Austria	Wagner.c@utanet.at
VOKS	Netherlands	Info@voks.nl
NFO	Norway	www.nfoe.homepage.com
Esofagus Gruppen	Sweden	Rmt@magotarm.sw
KEKS	Switzerland	Keka@datacom.ch

colon or stomach many surgeons have left the distal esophageal remnant in situ. However, it is now recognized that gastric mucosa can replace the normal squamous epithelium in these esophageal remnants, resulting in chronic inflammation and a Barrett's esophagus.[179] It is for this reason that it is recommended that the esophageal remnant should be completely excised, ideally at the time of esophageal replacement, or later if there is radiological evidence of esophagitis or ulceration, or if symptoms occur.[179,180]

A technique that used to be employed to gain gastrointestinal continuity following cervical esophagostomy involved creating an antithoracic tubularized bipedicle skin flap.[181,182] Squamous cell carcinoma is common in these conduits,[183] and they should be removed before adulthood.

At this stage we do not have accurate information on the relative risk of developing an esophageal malignancy after esophageal atresia repair, but it is likely to be increased. Clinicians need to be aware of the possibility that these patients may develop esophageal malignancy at a young age. It is yet to be determined whether the availability of proton pump inhibitors reduces this risk. In older patients, the role of regular endoscopy to assess the esophageal epithelium is yet to be established.

Quality of Life

Almost all adult patients enjoy a normal lifestyle, comparable to that of healthy adults in the general population.[168,169,184] Studies have not identified any differences in overall physical and mental health, and perhaps surprisingly, concomitant congenital abnormalities have not been found to influence generic quality of life. However, about one-third of patients report that the esophageal atresia had some negative effects on their lives, predominantly related to dysphagia (23%).[184]

Parent Support Groups

Details of some of the parent support groups for families who have a child with esophageal atresia are shown in Table 22.10. Many of these groups commenced in the 1980s. Some of them have extended their influence well beyond their own borders. For example, the German group KEKS has helped families in a number of war-torn countries and has brought families and babies born with esophageal atresia to Europe and cared for them in KEKS houses.

The International Esophageal Atresia team is made up of parents of esophageal atresia survivors who have children born with esophageal atresia and or tracheoesophageal fistula. They have joined together with other groups from around the world to offer information and support to families who are affected by esophageal atresia and the VATER association. Support groups can alleviate the isolation parents may feel,[168] and can provide practical assistance and emotional support to families in need. Some groups, e.g., AORA and CHEW, have been influential in setting up a number of major research studies.

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