CHAPTER

5 CONGENITAL ESOPHAGEAL ANOMALIES AND DIAPHRAGMATIC HERNIAS

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CONGENITAL ESOPHAGEAL ANOMALIES

EPIDEMIOLOGY

Esophageal atresia

Esophageal atresia occurs in about 2.4 per 10,000 live births,[1] with a slight preponderance in males and children of older or diabetic mothers.[2] Usually sporadic, familial patterns and genomic linkages have been observed. Children born to an affected parent have a 3 to 4 percent risk, children with one affected sibling have a 0.5 to 2 percent risk, and children with two affected siblings have a risk in excess of 20 percent.[3] Associated anomalies include cardiac malformations, gastrointestinal anomalies (e.g., anal atresia), urinary tract anomalies (e.g., uni- or bilateral renal agenesis or hypoplasia, multicystic kidney, horseshoe kidney), neural tube defects, and skeletal malformations.

Esophageal duplication cysts

Esophageal duplication cysts account for 10 to 15 percent of all foregut duplication cysts and only 5 to 10 percent of all mediastinal cysts.[4]

KEY CONCEPTS

- Epidemiology
  - Esophageal atresia occurs in about 2.4 of every 10,000 live births, with a slight preponderance in males and children of older or diabetic mothers. Esophageal duplication cysts account for 10 to 15 percent of all foregut duplication cysts and only 5 to 10 percent of all mediastinal cysts. Congenital esophageal stenosis occurs in between 1:25,000 and 1:50,000 live births. Bochdalek posterolateral diaphragmatic hernias occur in 1:2000 to 1:5000 live births. Morgagni retrosternal diaphragmatic hernias comprise 1 to 5 percent of congenital diaphragmatic defects, occurring much less frequently than Bochdalek hernias.
- Pathophysiology
  - Esophageal atresia constitutes a spectrum of incomplete esophageal development, sometimes coupled with communication to the tracheobronchial tree. This can result in discontinuity between the oropharynx and stomach, leading to feeding difficulties and a predisposition for respiratory infections. Esophageal duplication cysts can occur all along the esophagus, can be intramural, partitioned completely from the esophageal true lumen, or may communicate with it. Esophageal stenosis and webs can partially or completely obstruct the esophageal lumen. Much of the morbidity associated with these esophageal malformations is derived from a spectrum of associated malformations (i.e., cardiac, renal, neurologic, skeletal). Bochdalek diaphragmatic hernias result in displacement of abdominal viscera into a pleural space (usually the left side), leading maldevelopment of the ipsilateral lung; the associated contralateral mediastinal shift can also affect contralateral lung development. The end result is hypoxia and varying degrees of pulmonary hypertension. Morgagni hernias result in herniation of abdominal viscera into the mediastinum and can cause intestinal obstructive pathophysiology.

- Clinical features
  - Esophageal atresia is suggested by polyhydramnios coupled with a small or absent stomach in utero and excessive drooling, rhonchi, aspiration, and respiratory distress during initial postnatal feedings. Esophageal duplication cysts are often asymptomatic but can cause dysphagia or respiratory compromise secondary to mass effects. Congenital esophageal stenosis and webs typically present in early infancy, leading to progressive dysphagia and vomiting with ingestion of semisolid or solid foods. In its most severe presentation, newborns with Bochdalek hernias present with severe respiratory distress with a scaphoid abdomen. Morgagni hernias usually present much later in life with more subtle gastrointestinal symptoms or discomfort.
- Diagnostics
  - Esophageal atresia is most readily diagnosed with passage and insufflation of an esophageal catheter into the blind esophageal pouch. This can be imaged with plain film radiography; abdominal air confirms a distal fistula, whereas its absence suggests pure atresia. Plain films can also suggest esophageal cysts as sharply defined, spherical or tubular masses with esophageal and/or tracheal displacement. Confirmatory studies include contrast esophagography or computed tomography. Esophageal stenosis and webs are usually adequately diagnosed with barium esophagography and endoscopy. Plain chest radiography is often diagnostic for Bochdalek and Morgagni hernias but these can be confirmed with computed tomography, magnetic resonance imaging, or ultrasound.
- Treatment
  - Operative repair of esophageal atresia nominally consists of primarily anastomosing the proximal and distal esophageal segments with ligation of associated tracheoesophageal fistulas. Esophageal cysts are generally excised. Esophageal stenosis is usually initially treated conservatively with one or several dilations. Failing conservative therapy, surgical treatment generally consists of resecting the stenotic esophageal segment with primary anastomosis. Operative repair of Bochdalek and Morgagni hernias generally consists of reducing the herniated abdominal contents to their normal intraabdominal positions, resecting of any hernia sac, and primary or patch repair of the diaphragmatic defect.
- Results/outcomes
  - Repairs of esophageal atresia and stenosis are generally associated with good results; hospital survival rates range from 85 to 95 percent. Potential complications include anastomotic leak or stricture, recurrent fistula, and gastroesophageal reflux. Surgical repair of congenital diaphragmatic hernias is associated with good results. Morbidity and mortality with Bochdalek repairs usually arise from severe pulmonary developmental compromise and/or associated malformations.

Esophageal stenosis and webs

The incidence of congenital esophageal stenosis is reported to be between 1:25,000 and 1:50,000 live births.[5] Associated anomalies include esophageal or intestinal atresia, anorectal malformations, cardiac malformations, midgut malrotation, hypospadias, chromosomal abnormalities, and malformations of the head, face, and limbs.

Embryology

Esophageal atresia

Although the embryologic basis of esophageal atresia with or without tracheoesophageal fistula is poorly understood, it is postulated that this anomaly arises from failure of early endoderm-mesoderm interactions, which normally initiate development of the tracheobronchial tree from the esophagus, leading to "trachealization" of the proximal foregut with the main bronchi branching directly from this structure and the foregut continuing caudally to the stomach.

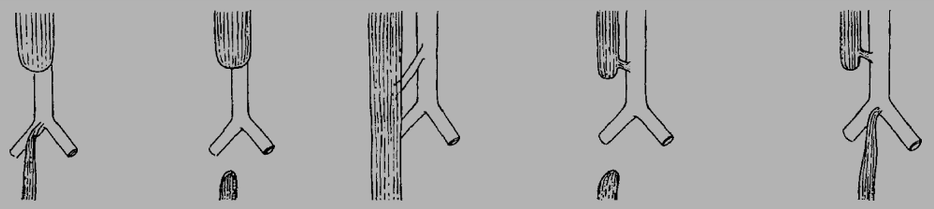
Esophageal duplication cysts

Several theories have arisen pertaining to the embryologic origins of esophageal duplication cysts. Kirwan and colleagues have suggested that errors in epithelial cell vacuolization during esophageal luminal development in the fourth through sixth weeks of gestation may be responsible.[6] Others implicate improper tracheobronchial budding.[7] The *endoderm-ectoderm adhesion theory* proposes that discordant longitudinal growth of the neural tube and foregut creates a shear force against adhesions that normally occur between the ectodermal and endodermal layers during primitive foregut development, detaching developing enteric cells.[8] The *split-notochord theory* postulates that esophageal cysts arise when an endodermic diverticulum expands posteriorly to fill an abnormal fissure between the endodermal and ectodermal layers of the primitive foregut.

Esophageal stenosis and webs

Fibromuscular thickening is the most common cause of congenital esophageal stenosis: however, its embryologic basis remains unknown. A less common cause of stenosis is tracheobronchial remnants, which are believed to result from incomplete separation of the primitive foregut from the respiratory tract. Congenital webs are thought to occur by failure of complete vacuolization of

Table 5-1 Gross and Ladd classification schemes for esophageal atresia and tracheoesophageal fistulas



Description	EA with distal TEF	Isolated EA	"N-type" TEF	EA with proximal TEF	EA with proximal and distal TEF
Gross Classification	C	A	E	B	D
Ladd Classification	III/IV[a]	I		II	V
Frequency (%)	86.5	7.7	4.2	0.8	0.7

EA = esophageal atresia; TEF = tracheoesophageal fistula.
[a]Type III fistula enters above tracheal bifurcation; type IV fistula enters at carina.
Source: Goldstein AM, Doody DP, Donahoe PK. Esophageal atresia and tracheoesophageal fistula. In: Pearson FG, Cooper JD, Deslauriers J, et al (eds). *Esophageal Surgery.* New York: Churchill Livingstone, 2002. With permission.

the mucosa-filled primordial esophageal lumen between the sixth and tenth weeks of embryogenesis.[6]

Classification

Esophageal atresia

The anatomic variants of esophageal atresia have been classified according to several different schemes. These variants generally comprise isolated esophageal atresia, esophageal atresia with a distal tracheoesophageal fistula, esophageal atresia with a proximal tracheoesophageal fistula, esophageal atresia with proximal and distal tracheoesophageal fistulas, and a so-called N-type tracheoesophageal fistula. The most commonly used classifications are those of Ladd[9] and Gross,[10] summarized in Table 5-1.

Esophageal duplication cysts

Esophageal cysts have been observed along all points of the esophagus, including the cervical, thoracic, and abdominal segments.[11] They can be intramural, completely partitioned from the esophagus, or they may communicate with the esophagus or other abdominal viscera. The definitions and classifications of esophageal duplication cysts have varied; however, Fallon and associates[12] have classified these cysts according to histologic and embryologic features (Table 5-2).

Esophageal stenosis and webs

Like esophageal duplication cysts, esophageal stenosis and webs have been classified according to several confusing schemes. A common scheme is that described by Nihoul-Fekete and coworkers (Table 5-3).[5]

Associated congenital anomalies

Esophageal atresia

Approximately 50 percent of infants born with esophageal atresia with or without tracheoesophageal fistula have additional congenital anomalies. Associated anomalies are seen more frequently in infants with an N-type fistula and those weighing less than 2000 g. A

Table 5-2 Categories of esophageal cysts

Category	Anatomic description
Intramural esophageal cysts ("true duplication" cysts)	Exist within the esophageal wall and are lined with squamous or columnar epithelium.
Enteric cysts	Circumscribed by well-developed muscular walls and contain epithelia from different embryonic origins.
Tracheobronchial foregut duplications	Anteriorly located cysts, most likely arising from a primitive lung bud that has incompletely separated from the primitive foregut. Lined with ciliated columnar or respiratory epithelium.
Posterior cysts	Often joined to the spinal column in the posterior mediastinum.
Neurenteric cysts	A subset of dorsal enteric cysts attached to the dura through a vertebral defect, often associated with vertebral anomalies (e.g., spina bifida occulta, anterior hemivertebrae).

Table 5-3 Categories of congenital esophageal stenosis

Category	Anatomic description
Fibromuscular thickening	Diffuse fibrosis of the wall with segmental hypertrophy of the muscularis and submucosal layers. Usually located in the distal esophagus, this is the type most commonly associated with esophageal atresia.
Tracheobronchial remnants	Comprising cartilage, respiratory mucous glands, or ciliated epithelium and forming a rigid, focal stenosis; most commonly located in the distal esophagus.
Membranous web	Consists of a thin, diaphragm-like membrane lined with epithelium and with an eccentrically located opening; observed at all esophageal levels. This is the rarest type of congenital esophageal stenosis.

thorough investigation into these associated anomalies is important, as they are often responsible for the morbidity and mortality seen in these patients.

Approximately 20 to 30 percent of infants with esophageal atresia have associated cardiovascular malformations,[13] the most frequently encountered anomalies associated with esophageal atresia. The most common of these include atrial and ventricular septal defects, patent ductus arteriosus, tetralogy of Fallot, and aortic arch anomalies. Gastrointestinal anomalies occur in about 25 percent of patients with esophageal atresia; the most commonly observed anomaly is anal atresia, followed by duodenal and ileal atresia, malrotation, Meckel's diverticulum, annular pancreas, and pyloric stenosis. Fortunately, most of these anomalies are comparatively easy to repair at the time of esophageal repair. Urinary tract malformations have been observed in as many as 24 percent of patients with esophageal atresia.[14] These anomalies include uni- or bilateral renal agenesis or hypoplasia, multicystic kidney, horseshoe kidney, and vesicoureteral reflux. Permanent renal dysfunction can be avoided if such anomalies are discovered and corrected early. Neural tube defects, hydrocephalus, and vertebral or extremity anomalies comprise neurologic and skeletal anomalies associated with esophageal atresia, observed in approximately 10 percent of affected infants.[15]

Esophageal atresia is one of the components of the VACTERL syndrome (i.e., vertebral anomalies, anal atresia, cardiac anomalies, tracheoesophageal fistula with esophageal atresia, renal defects, and radial limb dysplasia).[16] VACTERL occurs in approximately 15 percent of children with esophageal atresia, resulting in heightened mortality primarily due to cardiac malformations.

Esophageal duplication cysts

Esophageal cysts are associated with esophageal atresia,[17] vertebral anomalies, and neural tube defects.

Esophageal stenosis and webs

Congenital esophageal stenosis is associated with intestinal atresia, midgut malrotation, anorectal malformations, cardiac anomalies, hypospadias, chromosomal abnormalities, and malformations of the head, face, and limbs.[5] The fibromuscular thickening and tracheobronchial remnant types of esophageal stenosis are associated with esophageal atresia.

Clinical presentation and diagnosis

Esophageal atresia and tracheoesophageal fistula

Polyhydramnios coupled with a small or absent stomach observed on prenatal ultrasound examinations suggests esophageal atresia.[18] Pure atresia nearly always results in polyhydramnios, since polyhydramnios arises from an inability of the fetus to swallow amniotic fluid through an atretic esophagus. In cases of esophageal atresia without polyhydramnios, a distal tracheoesophageal fistula often permits the amniotic fluid to pass into the stomach.

In the early postnatal period, infants with esophageal atresia suffer from excessive drooling, accumulation of saliva in the posterior pharynx, excessive salivation from the nose and mouth, and rhonchi. Left undetected, this condition often leads to aspiration, with choking, respiratory distress, and cyanosis during the initial feedings; serious aspiration may also be associated with apnea, bradycardia, and death. A distal tracheoesophageal fistula predisposes to more severe respiratory distress, since gastric secretions can reflux into the tracheobronchial tree, causing pneumonitis and pulmonary sepsis. Other signs of pure esophageal atresia include a scaphoid abdomen. Atresia associated with a distal fistula can lead to a distended abdomen as air enters the stomach from the trachea. A complete physical exam can reveal or suggest other associated anomalies. Screening tests should include an echocardiogram, renal ultrasound, voiding cystourogram, and chromosomal analysis.

The diagnosis and delineation of esophageal atresia usually begins with passage of an esophageal catheter (e.g., 10F) through the infant's mouth. With slight insufflation of the tube, plain radiography can outline the size and shape of the esophageal pouch. Infrequently, dilute barium can be used to reveal a proximal tracheoesophageal fistula. On plain films, abdominal air confirms a distal fistula, whereas its absence suggests pure atresia. Other information obtained from the plain film includes signs of pneumonitis, an abnormal cardiac silhouette suggestive of a congenital malformation or right-sided aortic arch, and skeletal malformations.

An isolated N-type tracheoesophageal fistula usually leads to coughing and choking with feeding, gastric reflux into the tracheobronchial tree, and a barking cough due to tracheomalacia. Although contrast esophagography can be used to make this diagnosis, bronchoscopy and esophagoscopy are often required.

Esophageal duplication cysts

Esophageal cysts are often detected incidentally on plain chest radiographs, since many are asymptomatic. Symptoms arising from esophageal cysts usually consist of varying degrees of dysphagia. Esophageal cysts can also lead to respiratory distress in young patients, due to reductions in ventilatory volume, tracheal compression, or extrinsic compression of large bronchi, causing emphysema from air trapping and atelectasis or consolidation from extensive alveolar collapse. Other symptoms arise from the gastric epithelial cyst lining, including esophageal perforation, hemorrhage, or ulcerative pain. Rarely, larger cysts have elicited superior vena cava syndrome and abdominal masses. Neurenteric cysts commonly cause pain. Weakness and paralysis can arise from spinal cord compression caused by intraspinal lesions.

On plain chest radiography, esophageal cysts are usually suggested by a large unilateral (propensity to the right hemithorax), sharply defined, spherical or tubular mass. Often, this mass displaces the trachea and/or esophagus. Other plain film findings may derive from associated anomalies, including vertebral anomalies (e.g., bifid vertebrae). Confirmatory studies include contrast esophagography, typically demonstrating a smooth filling defect with luminal distortion or displacement. Transthoracic ultrasonography can also be used to confirm these lesions, differentiating them by their cystic nature from solid tumors. Computed tomography is replacing ultrasonography as the imaging modality of choice. Technetium scanning can reveal cysts containing a gastric mucosal lining, particularly in patients presenting with bleeding or ulceration. Esophagoscopy cannot reliably differentiate esophageal cysts from other posterior or mediastinal masses; however, this modality may be useful in identifying continuity between a cyst and the true esophageal lumen. Magnetic resonance imaging is useful in identifying any neurenteric components to esophageal cysts as well as detecting associated vertebral and intraspinal abnormalities.

Esophageal stenosis and webs

The typical presentation of congenital esophageal stenosis occurs in early infancy as progressive dysphagia and vomiting with ingestion of semisolid or solid foods. The severity of these symptoms largely depends on the degree of stenosis and its location within the esophagus; complete obstruction resembles esophageal atresia, proximal stenosis leads to an inability to swallow food, and distal lesions often result in regurgitation.

Diagnostically, barium esophagography and endoscopy are usually adequate in defining the location and severity of esophageal stenosis. Fibromuscular thickening is typically characterized by a long, tapered narrowing in the distal esophagus. Discrete, focal narrowings are more suggestive of tracheobronchial remnants. Webs or fibromuscular hypertrophy can also present as lesions in the middle or proximal third of the esophagus. Esophagoscopy usually demonstrates normal mucosa overlying a narrowed lumen; biopsy can exclude esophagitis and neoplastic lesions.

Preoperative management

Esophageal atresia and tracheoesophageal fistula

Infants with esophageal atresia and tracheoesophageal fistula should be nursed in a semiupright sitting position to reduce gastric reflux through the fistula into the tracheobronchial tree. Frequent oropharyngeal suctioning and a soft sump catheter placed into the atretic esophageal pouch can minimize aspiration events. Usually, prophylactic or therapeutic parenteral antibiotics (e.g., ampicillin and gentamicin) should be administered due to the high incidence of aspiration pneumonitis.

For infants suffering from severe aspiration pneumonitis or who are in significant respiratory distress, endotracheal intubation and mechanical ventilation is indicated. A gastrostomy should be considered to prevent continued reflux of gastric contents into the tracheobronchial tree and gastric distention. Early primary esophageal repair with division of an associated fistula can usually be performed within the first 2 days of life in stable patients without a major cardiac anomaly or respiratory compromise. Unstable patients with severe associated anomalies can be temporized with a gastrostomy until they have been optimized. In patients with congenital cardiac malformations, early primary esophageal repair can proceed in those whose circulation is not dependent on a patent ductus arteriosus.[13] In duct-dependent infants, prostaglandin E may be used to permit early repair. Otherwise, corrective cardiac surgery may have to performed first, with fistula division and gastrostomy placement sometimes performed during the cardiac operation; definitive esophageal repair should be delayed in these ill infants.

Esophageal duplication cysts

In most cases, elective surgical excision of esophageal cysts can be performed after diagnosis. In the rare cases of particularly large cysts associated with respiratory compromise, percutaneous decompression may be necessary preoperatively.[19]

Esophageal stenosis and webs

Effective treatment of congenital esophageal stenosis and webs focuses on alleviation of symptoms and preservation of normal antireflux mechanisms. Initial treatment for most children consists of esophageal dilatation by bougienage. This approach consists of a series of antegrade

and retrograde dilatations with mercury-weighted tapered Maloney or Avaray bougies and is most successful in children with a thin esophageal web or mild fibromuscular thickening. This conservative, nonoperative approach is still associated with occasional complications, including esophageal leak and recurrent stenosis. Long-term results are generally good, although many patients require repeated dilatations to maintain adequate esophageal patency.[20]

Operative repair

Esophageal atresia and tracheoesophageal fistula

Bronchoscopy should be performed prior to any operative repair of esophageal atresia in order to identify all tracheoesophageal fistulas, characterize tracheomalacia, and correctly position the endotracheal tube distal to the fistula and above the carina.

The operative approach is through a right posterolateral thoracotomy running 1 cm below the scapular tip (except in the case of a right-sided aortic arch) and within the inframammary crease (Fig. 5-1). Entry is usually through the fourth intercostal space, although the third interspace may afford better access to higher proximal pouches. Approaching the esophagus along the retropleural plane can limit any postoperative leak to the retropleural space. This approach is conducted by bluntly peeling the pleura away from the chest wall from the apex of the chest to a level several interspaces below the incision and posterior to the mediastinum.

The vagus nerve is identified and carefully preserved, while the azygos vein is divided. The trachea and distal esophagus are exposed by retracting the mediastinal pleura anteriorly. The distal esophagus is then looped to control any tracheoesophageal air leak, particularly if this interferes with mechanical ventilation during the procedure. The proximal esophageal pouch is then dissected up into the neck, identifying any proximal fistula; this can be facilitated with insertion of a Bakes dilator and gentle pushing of the proximal esophagus toward the surgeon by the anesthesiologist. If the gap between the proximal pouch and distal esophagus is so large as to preclude a primary repair, circular myotomies can be made in the proximal pouch to extend its length distally. Up to three myotomies can be made safely, with each adding approximately 1 cm of length.

Any tracheoesophageal fistula is isolated and divided; the tracheal side is closed with an interrupted absorbable 5-0 suture. The distal esophagus is mobilized sufficiently to permit tension-free approximation to the upper pouch. The proximal pouch is then opened and an end-to-end single-layer anastomosis is constructed with interrupted 5-0 nonabsorbable sutures on the posterior wall, with the knots tied on the outside. A feeding tube is then passed through the nares into the stomach across the anastomosis, followed by completion of the anterior circumference of the anastomosis; the feeding tube is secured to the nares to prevent dislodgement. If the distance between the proximal pouch and the distal esophagus is particularly long or the infant is premature, a gastrostomy is placed prior to the thoracotomy and a nasogastric feeding tube is not passed.

After the anastomosis has been completed, a chest tube is placed in the immediate vicinity of the repair but is secured to the posterior chest wall to prevent direct contact with the anastomosis. The thoracotomy is then closed in standard fashion. If primary repair is not possible, the distal esophagus is oversewn and sutured to the prevertebral fascia. Delayed primary repair is then undertaken after daily dilatations of the proximal pouch for 1 to 2 months. In those rare cases where a primary repair cannot be performed, alternatives include esophageal replacement using a colon interposition graft,[21] jejunal interposition graft,[22] or gastric tube.[23] Operative results are generally good, with hospital survival rates ranging from 85 to 95 percent.[1] Morbidity and mortality usually stem from associated congenital or chromosomal defects.

Complications Complications after operative repair of esophageal atresia include anastomotic leak (15 to 20 percent), recurrent fistula (3 to 10 percent), anastomotic stricture (10 to 35), gastroesophageal reflux (55 to 82 percent), and tracheomalacia (10 to 20 percent). Anastomotic leaks are often heralded with saliva observed in the chest tube, confirmed by orally administered methylene blue dye, and localized by barium swallow. Since most leaks are small, initial conservative management with proximal esophageal suctioning, localized drainage, antibiotics, and parenteral nutrition is often successful. Larger leaks or those that do not seal after conservative management should be operatively repaired to prevent mediastinal sepsis or empyema.

Infants with recurrent tracheoesophageal fistulas usually present several months after primary repair with cyanosis, wheezing, coughing or choking with feeding; abdominal distention, and recurrent pneumonias. The initial diagnosis is generally a barium swallow to identify and localize these fistulas. Since most of them do not close with expectant management, operative repair should generally be undertaken. The operative approach is dictated by the location of the fistulas; a cervical approach is taken for proximal fistulas while more distal fistulas are accessed via the chest. The fistulas must be closed at either end, followed by interposition tissue flaps placed between the esophagus and trachea to prevent recurrence.

Esophageal anastomotic strictures are best characterized by barium swallow and usually effectively treated with balloon dilatation. Gastroesophageal reflux—accompanied by heartburn, dysphagia, chronic coughing, vomiting, and recurrent respiratory infections—is commonly encountered after esophageal atresia repair. Once confirmed with a swallow study, symptomatic patients are initially treated conservatively with antacid therapy, thickened feeds, upright feedings, and promotility

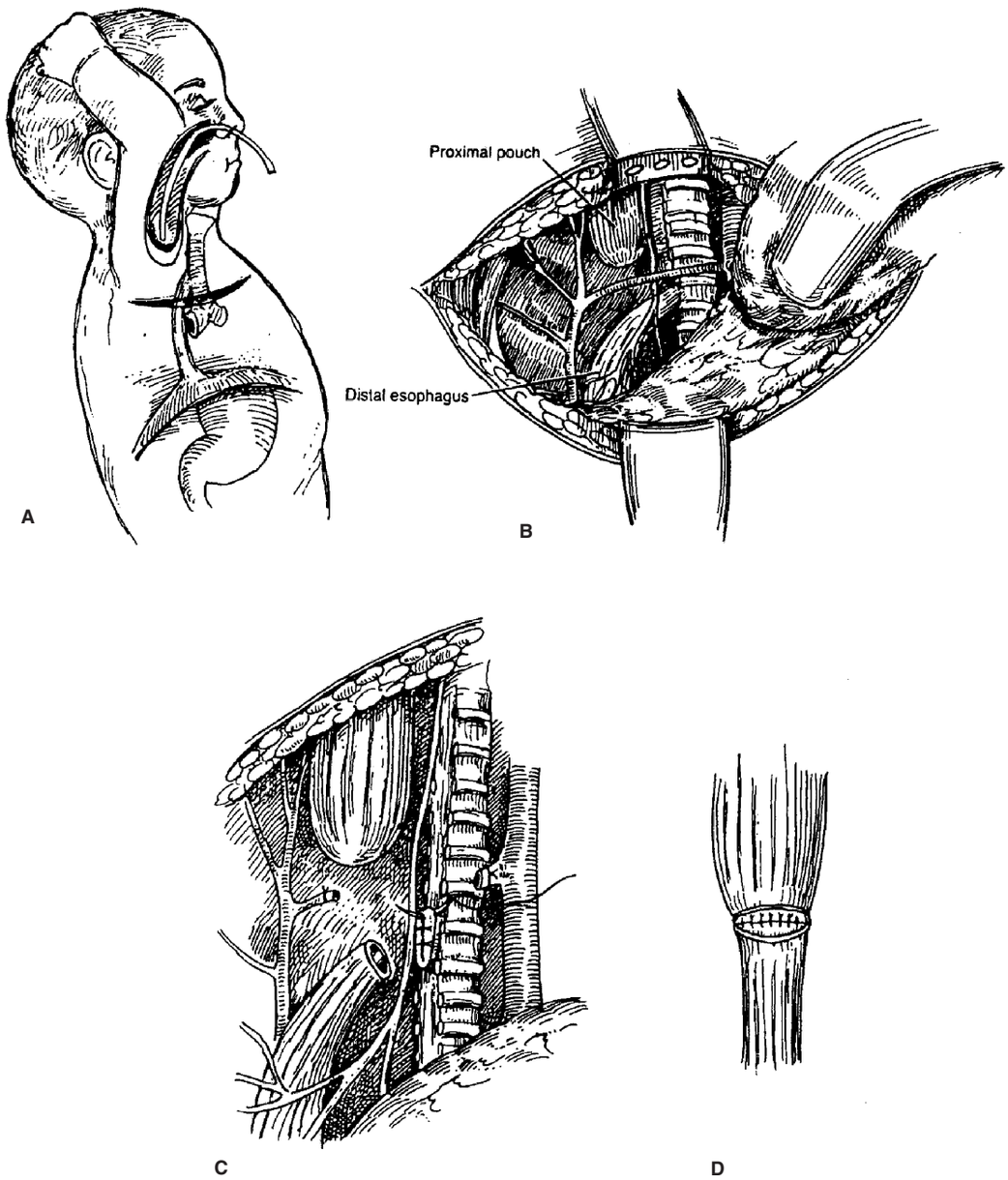


Figure 5-1 Surgical repair of type I esophageal atresia. A. A right posterolateral thoracotomy through the fourth intercostal space is a standard approach. B. The proximal esophageal pouch and distal esophagus are exposed with anterior traction of the lung and mediastinal pleura. C. Division and closure of the tracheoesophageal fistula. D. Single-layer primary anastomosis between the proximal and distal esophageal segments using interrupted absorbable sutures. (From O'Neill JA. *Operative Surgery, Principles and Techniques,* 3d ed. Philadelphia: Saunders, 1990:1072. With permission.)

agents. An antireflux procedure (e.g., partial Thal fundoplication) is considered for patients refractory to these medical measures.

Tracheomalacia, occurring in approximately 15 percent of patients after esophageal atresia repair, presents with a range of symptoms from mild expiratory stridor and barking cough to more severe cyanosis, apnea, and bradycardia. Bronchoscopy is diagnostic. Although mild symptoms often respond to expectant management, more severe symptoms require operative intervention, usually in the form of an aortopexy performed through a left thoracotomy. With this procedure, the aortic arch is sutured to the posterior sternal table to prevent its compression of the weakened tracheal wall.

Esophageal duplication cysts

Esophageal cysts are generally approached and excised through a limited posterolateral thoracotomy. Intramural cysts can be enucleated by dividing the overlying esophageal musculature, shelling out the cyst extramucosally, and maintaining esophageal wall integrity.[24] In all cases it is necessary to completely excise the cyst mucosa. Any continuity between the cyst and the esophageal lumen should be closed and the muscular esophageal wall reapproximated without creating a stricture. Operative results are generally excellent.

Esophageal stenosis and webs

If conservative management of esophageal stenosis or webs with one or more dilatations proves unsuccessful, resection with primary anastomosis should be considered. Once the stenosis has been localized, it is generally approached via a right thoracotomy for middle-third lesions or a left thoracotomy for distal esophageal lesions. Most stenoses can be resected with construction of a primary end-to-end anastomosis. Rare cases of more extensive esophageal resections may require esophageal replacement with interposition grafts. Resected stenoses near the gastroesophageal junction usually prompt a concomitant antireflux procedure to avoid significant reflux.

Complications Complications of surgical resection include anastomotic leak and gastroesophageal reflux.

An alternative to extensive resection of long fibromuscular esophageal stenoses is myotomy, although the intermediate- and long-term results are indeterminate.

CONGENITAL DIAPHRAGMATIC HERNIAS

EPIDEMIOLOGY

Posterolateral hiatal hernia (Bochdalek)

Congenital posterolateral diaphragmatic hernias occur in 1:2000 to 1:5000 live births.[25] As with congenital esophageal anomalies, associated defects are frequent, occurring in approximately 40 percent of affected patients. Associated anomalies include cardiac, renal, neural, and gastrointestinal defects (e.g., malrotation).

Retrosternal anterior diaphragmatic hernia (Morgagni)

Congenital retrosternal anterior diaphragmatic hernias comprise 1 to 5 percent of congenital diaphragmatic defects, occurring much less frequently than Bochdalek hernias. These hernias present most commonly in adulthood, with the onset of symptoms. They have a predisposition to affect obese patients and women.

Pathophysiology

Posterolateral hiatal hernia (Bochdalek)

Bochdalek hernias are caused by failure of the pleuroperitoneal canal to close during the eighth gestational week. Some 80 percent of these hernias occur on the left side. The primary pathophysiologic processes are incited by the herniation of abdominal organs into the chest through the diaphragmatic defect, compressing and impeding the growth and development of the ipsilateral lung. Mediastinal shift to the contralateral side may affect the other lung as well. A true hernial sac is encountered in only 10 percent of cases. Developmental morphologic and biochemical defects have also been observed in affected infants. These infants are susceptible to bronchopulmonary dysplasia, possibly related to defective antioxidant and surfactant mechanisms; pulmonary hypoplasia; and restrictive pulmonary physiology. Pulmonary hypoplasia and pulmonary hypertension lead to varying degrees of hypoxia and right-to-left shunting, with a heightened reactivity of these abnormal lungs to stimuli that increase pulmonary vascular resistance (i.e., hypoxia, acidosis, hypercarbia, hypothermia).

Retrosternal anterior diaphragmatic hernia (Morgagni)

Morgagni hernias are located between the xiphoid and costochondral diaphragmatic attachments and are predominantly on the right side, as the pericardium protects the left side. They are thought to arise from the failure of the transverse septum to fuse to the sternum, resulting in a triangular defect.[26] Unlike the Bochdalek hernia, a true hernial sac is usually present when abdominal viscera protrude into the mediastinum.

Diagnosis

Posterolateral hiatal hernia (Bochdalek)

Early presenting symptoms generally comprise varying degrees of respiratory compromise. The most extreme presentation is that of a newborn who is markedly dyspneic, tachycardic, and cyanotic with a scaphoid abdomen. There are signs of contralateral mediastinal and tracheal shift, with decreased breath sounds on the ipsilateral side. Plain chest radiography is often diagnostic, revealing bowel loops on the ipsilateral side, contralateral mediastinal shift, and a paucity of abdominal bowel gas patterns. The tip of any naso- or orogastric feeding tube placed may be seen above the diaphragm in the chest. Infants presenting later in life often present with feeding difficulties, colic, and growth retardation, generally depending on the size of the defect and the extent of visceral herniation into the pleural space.

Retrosternal anterior diaphragmatic hernia (Morgagni)

Symptoms vary; however, crampy abdominal pain or obstructive signs and symptoms constitute most of these. Often, plain chest radiographs display foramen of Morgagni hernias as excessively large densities in the region of the pericardial fat pad.[27] Confirmatory imaging includes computed tomography, magnetic resonance imaging, and/or ultrasonography, which show abdominal viscera within the hernial sac. In some cases, herniation of omentum only can make the diagnosis more difficult.

Preoperative management

Posterolateral hiatal hernia (Bochdalek)

Newborns presenting with respiratory distress and a clinical suspicion of a Bochdalek hernia should undergo endotracheal intubation with mechanical ventilation, paralysis/sedation, and orogastric tube decompression of the gastrointestinal tract to head off visceral distention and further respiratory embarrassment. Initial ventilatory parameters should include an inspired oxygen fraction of 100 percent, peak inspiratory pressure under 30 cmH_2O, positive end-expiratory pressure less than 5 cmH_2O, and hyperventilatory frequency; high-frequency ventilation may be useful in some settings. In order to reduce vasoreactive pulmonary hypertension, the postductal arterial oxygen tension should be maintained above 100 mmHg, carbon dioxide tension below 30 mmHg, and pH above 7.50. Tromethamine or sodium bicarbonate can be used to treat acidosis. Systolic blood pressures should be maintained above 50 mmHg, although intravenous fluids should be used sparingly. After adequate resuscitation, most of these patients should be referred for expeditious surgical repair. In patients who cannot be adequately ventilated mechanically, extracorporeal membrane oxygenation (ECMO) can be employed.

Retrosternal anterior diaphragmatic hernia (Morgagni)

Once diagnosed, patients with Morgagni diaphragmatic hernias are prepared for operative repair in a routine manner.

Operative repair

Posterolateral hiatal hernia (Bochdalek)

Most left-sided Bochdalek hernias are repaired via a transabdominal subcostal approach. Right-sided hernias are usually approached transthoracically. Herniated viscera are returned to their normal anatomic positions; any hernial sac is dissected free and resected. The lung tissue should be inspected; however, no attempt at expanding a hypoplastic lung should be made. Occasionally, extralobar pulmonary sequestration is encountered; it should be resected at this time. Once the edges of the diaphragmatic defect are defined, repair can usually be effected primarily with interrupted nonabsorbable suture material (e.g., polypropylene) in a horizontal mattress pattern. Large defects or defects whose edges are under tension should be closed with synthetic patch material (e.g., Teflon, Dacron). With these repairs, the patch is fashioned and sewn to the edges of the diaphragmatic defect with horizontal nonabsorbable sutures with or without a running buttressing suture line. Bilateral chest tubes should be placed and connected to a water seal. If the patient is stable, malrotation should be repaired at this time. In some cases, the abdominal wall fascia cannot be closed primarily. These cases are managed in similar fashion to large abdominal wall defects. Survivors of this operation generally have a good long-term prognosis, although some respiratory abnormalities have been observed in some survivors.

Retrosternal anterior diaphragmatic hernia (Morgagni)

Morgagni hernias are usually approached transperitoneally through an upper abdominal incision. The patient's habitus and size and the location of the hernial sac and its contents determine whether a subcostal, paramedian, or midline incision is used. The basic principles of hernia repair are used. Once the peritoneum is entered, the abdominal viscera are retracted from the hernial sac and returned to their normal locations. The hernial sac is then defined, taking down any associated adhesions, and resected. Small diaphragmatic defects that can be closed without tension can be repaired with nonabsorbable suture material (e.g., polypropylene), using a horizontal mattress pattern with or without a running reinforcement. Large defects or defects whose edges are under tension should be closed with synthetic patch material (e.g., Teflon, Dacron). With these repairs, the patch is fashioned and sewn to the edges of the diaphragmatic defect with horizontal nonabsorbable sutures with or without a running buttressing suture line. Morgagni repairs can be effected via a thoracic approach. In these cases the principles and techniques of repair resemble those used in the transperitoneal approach. The results with these repairs are generally excellent.

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