Pediatric Chest I
Developmental and Physiologic Conditions for the Surgeon

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KEYWORDS
- Pediatric chest
- Pediatric mediastinum
- Pediatric thorax anatomy
- Pediatric thorax surgery

KEY POINTS
- The mediastinum, pleura, and lungs are a collection of complex organs interacting in a constant and literally fluid manner.
- A pneumothorax results from the separation of the visceral pleura of the lung from the parietal pleura of the chest wall.
- Surgical management of congenital and acquired conditions of the pediatric thorax is predicated on the understanding of anatomical and physiologic development, appropriate anatomical relationships, and appropriate surgical principles concerning the thorax.

This article addresses basic anatomic considerations of the chest and describes common conditions of the lungs, pleura, and mediastinum that affect children. Treatment of malignant conditions and management of congenital diaphragmatic hernia and congenital chest wall deformities are addressed in other articles by Ken Azarow elsewhere in this issue.

ANATOMY

The mediastinum, pleura, and lungs are a collection of complex organs interacting in a constant and literally fluid manner. The mediastinum comprises the central portion of the thoracic cavity. Its boundaries are the sternum anteriorly, the vertebral column posteriorly, and the medial parietal pleural surfaces of the right and left lungs laterally. The mediastinum is sometimes described as having 4 compartments: superior, anterior, middle, and posterior.\textsuperscript{1} However, clinically, lesions are usually described as being
within 1 of 3 compartments: anterior, middle, and posterior. In this schema, the superior compartment is subsumed into the other compartments. The anterior mediastinum is bounded by the sternum anteriorly, the diaphragm inferiorly, the trachea, great vessels, and pericardium posteriorly, and the pleurae and lungs laterally. The anterior compartment contains the thymus, lymphoid structures, and nerves and vessels. The middle mediastinum is bounded by the anterior and posterior limits of the pericardium. It contains the trachea, main-stem bronchi, the heart and great vessels, and hilar lymph nodes. The posterior mediastinum is bounded inferiorly by the diaphragm, anteriorly by the pericardium, and posteriorly by the vertebral column. It contains the aorta, thoracic esophagus, and the sympathetic nerve chain (Fig. 1).

The right and left pleural spaces are separate entities. They envelop each lung as they extend from the lateral aspects of the mediastinum. The parietal pleura lines the thoracic wall and is continuous with the visceral pleura as it adheres intimately to the pulmonary surface. A thin layer of mesothelial cells and fluid is interposed between the 2 surfaces as they adhere to each other. In some areas, the parietal pleura may fold onto itself until the lung intercedes during inspiration. These areas are found inferiorly along the edges of the diaphragm, in the costodiaphragmatic sinus, and in a small cleft behind the

Fig. 1. Anatomic division of the mediastinum: anterior compartment extends from the sternum to the dotted line anterior to the pericardium. Middle mediastinum extends posteriorly to the anterior border of the vertebrae (solid line). AV, azygous vein; IVC, inferior vena cava; LN, lymph node; PA, pulmonary artery; PV, pulmonary vein; RIV, right internal jugular vein; RMB, right main-stem bronchus; SVC, superior vena cava. (From Tovar JA. Mediastinal tumors. In: Holcomb III GW, Murphy JP, editors. Ashcraft's pediatric surgery. 5th edition. Philadelphia: Saunders; 2010. p. 322; with permission.)
sternum known as the costomediastinal sinus. The parietal pleura receives its blood supply from the intercostal, internal mammary, superior phrenic, and anterior mediastinal arteries. Corresponding veins drain the parietal pleura into the systemic veins. The visceral pleura receives its perfusion from bronchial and pulmonary artery radicals. Venous drainage is only to the pulmonary circulation. The parietal pleura receives sensory innervation from the intercostal and phrenic nerves, resulting in relatively precise sensory localization. The visceral pleura receives vagal and sympathetic innervation and has less precise sensory localization.1,3

The systemic and pulmonary vasculature and the lymphatic circulation interact to maintain a relatively constant amount of pleural fluid, which is evenly distributed within the pleural space.1,4,5 Most pleural fluid is formed from the systemic circulation. It then travels along pressure gradients in the pleural space until it is primarily resorbed by parietal pleural lymphatics. Increase of systemic venous pressure or lymphatic pressure results in excessive accumulation of pleural fluid either by increased production (venous congestion) or decreased absorption (lymphatic congestion). Normally, there is little interchange between pleural and pulmonary fluids. However, pulmonary edema can contribute to an increase in pleural fluid by presenting a greater amount of fluid to the visceral pleura from the pulmonary parenchyma itself.1,6

In children, the lungs and airways are not fully developed at birth. In the postnatal period, the number of immature alveoli continues to increase. They subsequently enlarge into mature alveoli. The area of the air-blood interface continues to increase as alveoli and capillaries multiply. These changes continue until at least the eighth postnatal year. Only about 50 million alveoli (one-sixth of the adult number) are present at birth. The structural integrity of the airway improves after birth as the flexible cartilage of the infant’s larynx and trachea becomes more rigid.1,7

PLEURA AND LUNGS

Pneumothorax

A pneumothorax results from the separation of the visceral pleura of the lung from the parietal pleura of the chest wall. Air accumulates in this space. If the air accumulates under pressure, a tension pneumothorax ensues. A pneumothorax may occur spontaneously or as the result of trauma, surgery, or a therapeutic intervention. Pulmonary volume, compliance, and diffusing capacity are compromised and if the pneumothorax is substantial, hypoxia may result secondary to ventilation-perfusion mismatch. A normal lung may be able to compensate for this situation. Children with underlying chronic pulmonary disease may suffer relatively smaller pneumothoraces secondary to diminished elastic recoil of their lungs, but the symptomatic consequences may be more significant because of their small margin of pulmonary reserve.8,9

Many pneumothoraces experienced by children are spontaneous. Most of these spontaneous pneumothoraces occur in the adolescent age group. The typically affected adolescent is 14 to 18 years old, male, tall, and thin. Few of these youngsters are smokers.10,11 Spontaneous pneumothorax often occurs in the absence of any underlying disease. However, it may result from an underlying condition such as a congenital or acquired bleb, familial tendency, pneumonia with pneumatocele or abscess, tuberculosis, pleuropulmonary blastoma, osteochondroma, or cystic adenomatoid malformation.8,12–17

Pneumothorax is also common in the infant population. Symptomatic pneumothorax is estimated to occur in 0.08% of all live births, and in 5% to 7% of infants with a birth weight 1500 g or less. Typical risks include respiratory distress syndrome, meconium aspiration, pulmonary hypoplasia, continuous positive airway pressure,
and mechanical ventilation. Risks may be lowered by the use of surfactant and synchronized and low-volume ventilation.\textsuperscript{18–21}

Pneumothorax from trauma may result from a tear in the pleura, esophagus, trachea, or bronchi. Iatrogenic causes include mechanical ventilation, thoracentesis, central venous catheter insertion, bronchoscopy, or cardiopulmonary resuscitation.\textsuperscript{8,12–14}

Spontaneous pneumothorax most commonly presents with chest pain. Accompanying symptoms may also include back or shoulder pain, dyspnea, and cough. In the trauma setting, pneumothorax is routinely screened for. But the presence of the signs and symptoms noted earlier should raise suspicion for pneumothorax. In any infant or child who is already being ventilated, any decline in respiratory status should also raise suspicion for pneumothorax.\textsuperscript{8,10,11,13,21}

Severe dyspnea or respiratory insufficiency should alert the provider to the presence of a tension pneumothorax. Physical examination of a patient with pneumothorax reveals diminished breath sounds on the affected side of the chest. The trachea may be shifted away from the side of a tension pneumothorax. Pneumothorax should be visible on a chest radiograph and is enhanced if the radiograph is taken at end expiration. It is common practice for the size of a pneumothorax to be described as a proportion of the chest field on an upright radiograph. However, the volume loss of the lung is greater than such a description because pulmonary volume is lost in 3 dimensions.

The factors that determine the proper management of pneumothorax include the initial size, signs and symptoms, ongoing expansion, presence of tension, and any contributing underlying condition. A spontaneous unilateral pneumothorax that is asymptomatic and less than 15% to 20% of the chest volume can usually be followed by observation alone.\textsuperscript{8,10,11,14} Pleural air reabsors at a rate of 1.25% per day, but this can be hastened by breathing supplemental oxygen.\textsuperscript{8,22} Options for treatment of a small pneumothorax include thoracentesis, placement of a Heimlich valve, or placement of a pigtail catheter.\textsuperscript{10,11,23–25} However, if air is continuously aspirated, the pneumothorax persists, symptoms are initially significant or not relieved by the initial maneuvers, or the presenting pneumothorax is large in size, a standard chest tube should be inserted.

A tension pneumothorax requires emergent placement of a chest tube. If a tube is not immediately available or if the patient’s condition deteriorates during preparation for placement, a large-bore (14-gauge) needle should be placed just over the rib in the anterior second intercostal space in the midclavicular line to relieve the tension. A chest tube can then be placed.

Pneumothoraces in infants, particularly infants being ventilated, may sometimes be treated expectantly by keeping ventilator settings low, by using an oscillatory ventilator, and by keeping the affected side down. Most commonly, needle aspiration or chest tube drainage is required. Neonates are most prone to injuries from chest tubes that can result in lung tears, phrenic nerve injury, chylothorax and pericardial effusions.\textsuperscript{21,26} Fibrin glue has been used in infants to try to seal pneumothoraces. However, this treatment carries risks of hypercalcemia, bradycardia, diaphragm paralysis, and skin necrosis.\textsuperscript{26,27}

In a posttraumatic pneumothorax, large or persistent air leaks may indicate damage to the airway or the esophagus. Appropriate diagnostic studies using an esophagram, bronchoscopy, esophagoscopy, thoracoscopy, or thoracotomy should be undertaken and direct repair of the injury, if present, performed. If the air leak is the result of lung parenchymal injury, chest tube drainage is usually corrective.

If a spontaneous pneumothorax occurs for the first time, initial treatment is usually limited to one of the types of chest drainage noted earlier. A recent study assessing
cost and morbidities of treatment options advocates against aggressive surgical intervention as a primary treatment of spontaneous pneumothorax. Pneumothorax recurs with a frequency of 50% to 60% after the first episode in children. If a persistent air leak is present, or if the pneumothorax recurs after removal of the chest tube, then further surgical intervention is warranted. Such intervention is now typically performed using a video-assisted thoracoscopic (VATS) technique.

Once a VATS procedure is undertaken, the goal is to remove any resectable lung disease if present and perform pleurodesis. Blebs and cysts can usually be removed with a stapled or ligated wedge resection. Fibrin glue or absorbable mesh, or both, are sometimes used to reinforce the resected area. Pleurodesis can be undertaken using an instilled agent, mechanical abrasion with a bovie pad or sponge, or both. The most commonly used pleurodesis agent is USP purified talc. A recent study in adults suggested that talc may have systemic distribution and may rarely produce acute respiratory distress syndrome (ARDS). However, multiple other reports have advocated the safety of talc. Treatment with talc has been shown to be particularly effective in treating pneumothoraces in children with cystic fibrosis. Other agents such as silver nitrate, quinacrine, iodized oil, minocycline, doxycycline, and hypertonic glucose have also been reported as potential agents, but are not commonly used. The results of VATS pleurodesis for pneumothorax have been excellent, and complications of the technique are uncommon.

Empyema

Empyema refers to the accumulation of infected fluid in the pleural space. In children, empyema is most commonly secondary to an underlying pneumonia. Other causes may include perforation of the esophagus or tracheobronchial tree, spontaneously or from trauma, and spread of infection from retropharyngeal, mediastinal, or paravertebral spaces. These latter causes are less common in children than in adults. In 1962, the American Thoracic Society described what are still considered the 3 classic stages of empyema. The first stage, or the exudative stage, lasts for about 24 to 72 hours. It is characterized by an accumulation of thin pleural fluid and is amenable to drainage by thoracentesis. The second stage is the fibrinopurulent stage. This stage lasts 7 to 10 days and is characterized by the formation of a thicker, more fibrinous fluid with loculations. The lung loses its mobility in this stage. The third stage is the organizing stage. This stage usually occurs about 2 to 4 weeks after the onset of the empyema. A pleural peel forms and the lung becomes entrapped.

Historically, the most common organisms causing empyema have varied. Streptococcus pyogenes, Staphylococcus aureus, and Haemophilus influenzae were most common before the advent of antibiotics. Subsequent offending organisms then emerged depending on the nature of antibiotics developed. Currently, Streptococcus pneumoniae along with other streptococcal species, and Staphylococcus aureus, including methicillin-resistant Staphylococcus aureus, are the leading causative organisms of empyema. The incidence of Streptococcus pneumoniae is increasing despite the use of pneumococcal vaccine. Children with empyema most commonly present with fever. Other symptoms include cough, respiratory insufficiency, chest pain, and sometimes abdominal pain. Physical signs may include dullness on chest percussion, tactile and vocal fremitus, decreased breath sounds, rales, and a pleural friction rub. A chest radiograph reveals a density on the affected side of the chest that reflects pleural fluid accumulation and thickening as well as parenchymal compression. In advanced empyema, there may be shifting of the mediastinum to the contralateral side, and there may be air-fluid levels in a loculated area, or in a lung abscess. Transthoracic ultrasonography
or a chest computed tomography (CT) scan can better determine the degree of pleural thickening, fluid loculation, and lung consolidation. The diagnosis of empyema is confirmed by thoracentesis. The fluid is characteristically turbid and may be thick during the later stages of the infection. Laboratory analysis reveals a specific gravity greater than 1.016, protein greater than 3 g/dL, lactate dehydrogenase (LDH) level greater than 200 U/L, pleural fluid protein/serum protein ratio greater than 0.5, pleural fluid LDH/serum LDH ratio greater than 0.6, and white blood cell count higher than 15,000/mm³. Fibrin clots may also be present.

Once the diagnosis of empyema is made, and if fluid is obtained before the initiation of treatment, antibiotic therapy can be targeted toward the offending organism. However, it is common in children for antibiotics to be started without a fluid sample. If fluid is obtained after this point, it is often unrevealing. It is possible to treat pneumonia with a small to moderate empyema with antibiotics alone. However, if a child is showing respiratory insufficiency, or if antibiotic treatment is not showing progress in resolving fever or symptoms such as pain within the first few days, then the empyema should be drained. As complete a drainage of the empyema as possible should be accomplished either by thoracentesis, tube thoracostomy with or without fibrinolytic agents, or thoracoscopic drainage. Thoracotomy is rarely needed for the treatment of empyema. In general, the longer the prehospital or pretreatment illness has persisted, the more involved the interventions that are needed may be.

There is debate within the literature about the optimal drainage procedure for empyema. The surgical objective is to remove as much of the infected pleural fluid and debris as possible, to break up any loculations, and to free the lung to expand and to fill the pleural space. The lung expansion allows better clearance of infection from the lung parenchyma itself. Unlike adults, children rarely develop any chronic constricting pleural rinds with appropriate therapy. Children usually need sedation or anesthesia for any type of invasive procedure such as thoracentesis or chest tube placement. This situation has led some investigators to advocate VATS-assisted drainage early in the patient’s course. It is believed that this strategy leads to a faster, more complete evacuation of the chest, a faster resolution of symptoms, and a decreased length of stay compared with other interventions. In addition, it may prevent multiple procedures in the case of failed initial therapy with thoracentesis or thoracostomy with or without fibrinolysis. Others believe that thoracostomy

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**Fig. 2.** (A) Empyema of right chest; density reflects pleural fluid accumulation, pleural thickening, and parenchymal compression. (B) CT image of right chest empyema; arrow denotes compressed lung parenchyma and pneumatoceles within lower lobe.
with fibrinolysis, using agents such as streptokinase, urokinase, or tissue plasminogen activator, is as effective as VATS drainage in most cases, and that it may be more cost-effective with less morbidity than VATS. However, only 1 prospective study of these 2 modes of therapy has been performed. A position paper by the American Pediatric Surgical Association in 2009 suggests an algorithm for treatment of empyema (Fig. 4). The algorithm indicates that simple empyemas present for 5 days or less may be treated with catheter drainage alone. Empyemas that are loculated or present for greater than 5 days may be initially addressed with VATS or with chest tube placement with fibrinolytic therapy. The need for a prospective study in this regard is acknowledged in this article.

Radiographic resolution of empyema typically lags behind clinical response. Therefore, persistent findings of radiographic abnormalities should not be an indication for repeat or further intervention if the child is clinically recovering. If a lung abscess develops, treatment may require pneumonostomy, wedge resection, or lobar resection. After a child has clinically recovered from an empyema, it is advisable to follow serial chest radiographs over time until the lung fields return to normal to be sure there were no previously unknown inciting causes of the infection such as congenital cystic adenomatoid malformation (CCAM) or sequestration. With prompt and appropriate therapy, the overall outcome for children with empyema is excellent. Pulmonary function after recovery is usually clinically normal. However, some investigators have found mild restrictive or obstructive disease on follow-up spirometry.

**Chylothorax**

Chylothorax is the accumulation of lymphatic fluid within the pleural space. Congenital chylothorax is reported to be the most common cause of pleural effusion in neonates. In this age group, it is believed to be caused by abnormalities in the development of the lymphatic system such as lymphangiomatosis or lymphangiectasia. It may be present prenatally on ultrasonography and is sometimes associated with fetal hydrops. Prenatal chylothorax is a lethal condition and may require prenatal intervention such as thoracoamniotic shunting. Another major cause of chylothorax in
Infants and children are injury to lymphatic channels as a result of an operative procedure, most typically for cardiac anomalies but also after repair of congenital diaphragmatic hernia, amongst other conditions. The incidence of chylothorax after thoracic surgery in children reportedly ranges from 0.25% to 0.9%.91–93 The incidence after repair of congenital diaphragmatic hernia is 5% to 10%.94–96 Other causes of chylothorax in children are malignancy, particularly neuroblastoma and lymphoma, occlusion or thrombosis of the upper central venous system, hypothyroidism, a variety of syndromes, and traumatic injury caused by either blunt or penetrating thoracic trauma.

Fig. 4. Proposed algorithm for the treatment of children with pneumonia and parapneumonic effusions or empyema. (From Kokoska ER, Chen MK, The New Technology Committee. Position paper on video-assisted thoracoscopic surgery as treatment of pediatric empyema. J Pediatr Surg 2009;44:292; with permission.)
or child abuse.\textsuperscript{87,97–100} If an older child presents with chylothorax but has no history of trauma or operation, an intrathoracic tumor should be suspected and investigated with chest CT or magnetic resonance imaging (MRI).

Chylothorax typically presents with respiratory insufficiency, although it may also be first recognized as the incidental finding of a pleural effusion on an imaging study of the chest. As noted earlier, it may also be found on prenatal ultrasonography. The diagnosis of chylothorax is confirmed by evaluating the pleural fluid. Chyle is usually milky, but may be serosanguinous or straw-colored in children who are receiving no enteral fats, such as those who have just undergone surgery. One recent report defines chylothorax as having the following characteristics: more than 1000 leukocytes/mL (>70% lymphocytes), triglycerides greater than 100 mg/dL; protein greater than 20 g/L; sterile culture, positive Sudan III (a fat-soluble dye) staining of the fluid from enterally fed patients.\textsuperscript{101} Loss of significant volumes of chyle from the pleural space results in a substantial loss of protein and lymphocytes. These losses must be monitored closely and replaced to avoid severe nutritional deficits in affected children.

Initial treatment of chylothorax is nonoperative, with about 70% to 80% of patients responding appropriately.\textsuperscript{101–104} The pleural space is drained by thoracentesis or placement of a chest tube to relieve symptoms and facilitate closure of the lymphatic leak. Administering a diet that principally contains medium-chain triglycerides can diminish lymphatic flow through the thoracic duct. These fats are absorbed directly into the portal venous system, unlike long chain fatty acids, which are absorbed through the intestinal lymphatics. If chyle drainage persists, the patient can be placed on total parental nutrition, with cessation of enteral intake. If these measures fail to significantly reduce or eliminate lymphatic drainage within a few days, administration of octreotide, an analogue of somatostatin, may be helpful. This procedure can also be used in conjunction with the interventions noted earlier. It is uncertain how octreotide inhibits lymphatic flow, but it may act directly on somatostatin receptors in the splanchnic circulation to reduce lymph fluid production and thereby reduce its passage through the thoracic duct. It may also reduce the volume and protein content of lymph fluid within the thoracic duct by reducing gastric, pancreatic, and bile secretions.\textsuperscript{87} No prospective study has been performed to evaluate the efficacy of somatostatin or octreotide in treating chylothorax.\textsuperscript{105} However, there are multiple reports of the efficacy of somatostatin and octreotide in this regard.\textsuperscript{87,102,104,106–110} Other reports indicate no particular success with these agents.\textsuperscript{92,111}

If nonoperative treatment fails to resolve a chylothorax, surgical intervention is indicated. Options include ligation of the thoracic duct, embolization of the thoracic duct, pleurodesis or pleurectomy, sealing the area of leakage with fibrin glue, or the use of a pleural-peritoneal shunt. Thoracotomy or thorascopic technique may be used when appropriate.\textsuperscript{112–119}

The timing of surgical intervention has been a matter of debate. Contemporary reports recommend intervention at as early as 5 to 7 days, particularly if less invasive procedures such as thoracoscopy or pleuroperitoneal shunts are to be used.\textsuperscript{115,118,120} This situation is especially true if the child has increased right-heart pressures or central venous thrombosis. Such children are unlikely to respond to nonoperative treatment.\textsuperscript{115}

\textbf{CCAM}

CCAM is a rare anomaly with potentially serious clinical implications. These lesions are also referred to as congenital pulmonary airway malformations. The terminology still seems unsettled in the literature and therefore the more traditional CCAM terminology is used in this section. The reported incidence of CCAM is between 1:25,000 and
1:35,000 live births and it is the most common congenital cystic lung lesion. These lesions are usually confined to a single lobe and are more commonly left sided (Fig. 5).\textsuperscript{121,122} Bilateral lesions are rare.\textsuperscript{123} There is an 18% rate of associated abnormalities, which include renal agenesis and cardiac anomalies.\textsuperscript{121} The differential diagnosis includes pulmonary sequestration, bronchogenic cyst, congenital lobar emphysema (CLE), enteric duplication, and congenital diaphragmatic hernia.\textsuperscript{124}

Embryologically, CCAMs are believed to arise from a derangement of lung matura-
tion at the level of the terminal bronchioles, leading to suppression of alveolar develop-
ment and disorganized proliferation, resulting in cyst formation.\textsuperscript{121–123} Although clinically distinct from pulmonary sequestrations, hybrid lesions do exist that contain features of both CCAM and sequestrations. These lesions usually meet criteria for CCAM but also have a supplemental systemic arterial supply. This discovery has led many investigators to suggest that CCAM and sequestrations may represent a spectrum of the same embryologic process.\textsuperscript{125}

CCAMs are generally classified by cyst size and histology. The Stocker classification system is the most commonly used means of description. Type 0 consists of small solid lungs and is incompatible with life. Type I (most common) consists of single or multiple cysts of greater than 1 cm in diameter lined with pseudostratified epithelium, mucus-secreting cells, and prominent cartilage.\textsuperscript{126} Type II is defined as having multiple cysts, all less than 1 cm in diameter, composed of cylindrical or cuboidal epithelium with prom- inent smooth or striated muscle. Type III is grossly solid but microcystic, lined by cuboidal epithelium and often intricately folded.\textsuperscript{125} Type IV consists of large cysts lined with flattened epithelium surrounded by loose mesenchymal tissue. Types I and IV are considered to have a favorable prognosis. The remaining types have a poorer prognosis because of increased rates of other associated anomalies.\textsuperscript{126}

The increasing use and sophistication of prenatal Doppler ultrasonography (DUS) has led to an increase in the reported diagnosis of CCAM. Lung malformations may be detected by DUS as early as 18 to 20 weeks’ gestational age with a sensitivity of 76% to 81%.\textsuperscript{122,125,127} Although definitive diagnosis of a specific lung lesion is uncertain on DUS, a CCAM typically appears as large macrocystic lesions or as a solid, echogenic-appearing mass. If a systemic feeding vessel is identified, then the mass is more likely a pulmonary sequestration or hybrid lesion.\textsuperscript{123} Some CCAMs may appear to regress or involute on serial DUS. Postnatal chest radiography may show a mass,

Fig. 5. (A) Chest radiograph depicts CCAM (arrows) in left lower lung field. (B) CT image shows the CCAM in (A) to involve the left lower lobe.
a cystic lesion, or hyperinflation, or may be normal.\textsuperscript{125,128} Even if a lesion appears to resolve on DUS, or is not seen on postnatal chest radiography, these lesions are usually identified with postnatal CT or MRI, which have been reported to have a 100\% specificity in these cases.\textsuperscript{122,123,125,129} Such imaging should be performed in all asymptomatic infants within the first 6 to 12 months of life, or sooner in symptomatic infants. This imaging is helpful in identifying the extent of the lesion for operative planning. There is little evidence to suggest that lesions present postnatally involute further.\textsuperscript{123,124} Although most patients are born with few or no symptoms from their lesion, CCAMs may grow large enough to cause significant impairment both prenatally and postnatally.\textsuperscript{122,128,129} Prenatally, the lesion may present with polyhydramnios, hydrothorax, or hydrops fetalis.\textsuperscript{123,129,130} Compression of the fetal esophagus by a large intrathoracic mass interferes with fetal swallowing and produces polyhydramnios. Cardiac and vena cava compression caused by significant mediastinal shift by a large CCAM may lead to hydrops fetalis. Adzick and colleagues\textsuperscript{123} have reported that a cystic adenomatoid volume ratio (CVR) greater than 1.6 is predictive of an increased risk of hydrops. The CVR is calculated by dividing CCAM volume by head circumference. Patients presenting with, or at risk for, development of hydrops may be considered for fetal intervention consisting of fetal pulmonary lobectomy or ex utero intrapartum therapy. Fetal hydrothorax may be amenable to thoracoamniotic shunting.\textsuperscript{123,124,130} Postnatally, patients may be asymptomatic or present with cough, respiratory distress, recurrent infection, pneumothorax, or malignant degeneration.\textsuperscript{122,125,127,128} Although rare, CCAMs have been associated with pulmonary malignancy.\textsuperscript{128} Pleuropulmonary blastoma, bronchioalveolar carcinoma, pulmonary rhabdomyosarcoma, and mesenchymoma have all been described as arising within CCAMs.\textsuperscript{122,125,127,128,131–133} Pleuropulmonary blastoma has been shown to present more commonly in infants and young children, whereas older children and adults are more often diagnosed with bronchioalveolar carcinoma.\textsuperscript{125,127,133} Type I CCAM is associated with bronchioalveolar carcinoma, whereas type IV shows histologic overlap with pleuropulmonary blastoma.\textsuperscript{126,134} The overall risk of malignant transformation seems to be 2\% to 4\%.\textsuperscript{132}

Patients presenting with acute respiratory distress shortly after birth should undergo urgent resection of the CCAM once stable. Acute treatment of severely symptomatic neonates may include emergent chest decompression with tube thoracostomy.\textsuperscript{124} Most investigators agree that asymptomatic patients should undergo surgical resection on an elective basis at around 3 to 12 months of age, although the rate of infection begins to increase after 7 months of age.\textsuperscript{121,127,129} Anatomic lobectomy via standard thoracotomy or with a thorascoscopic approach is the standard of care.\textsuperscript{122,124,133,135} Thoracoscopy may generally be better tolerated and may result in better postoperative pain control, shorter hospital stay, a lower respiratory complication rate, and a lower risk of later chest wall deformity than thoracotomy.\textsuperscript{124,135} Conversion from thorascoscopic to open resection is directly correlated with previous episodes of pneumonia as a result of the development of adhesions within the pleural space.\textsuperscript{135} Pulmonary segmentectomy may be performed but carries a risk of failing to remove all of the abnormal tissue, thereby leaving a residual risk of infection and malignancy.\textsuperscript{122,125,133} Early resection has the added benefit of allowing for compensatory lung growth and development.\textsuperscript{122,125,129} Resection is indicated both to prevent symptoms but also to avoid malignant degeneration.\textsuperscript{124,132,133} The long-term outcome for patients with completely resected CCAM is excellent.\textsuperscript{128,129}

**Pulmonary Sequestration**

Pulmonary sequestration is a rare congenital malformation of the respiratory tract. It is defined as a mass of nonfunctioning pulmonary tissue that lacks a definitive
connection to the tracheobronchial tree and has a systemic arterial supply of variable origin.\textsuperscript{136,137} Classically, these lesions are subdivided into 2 types: intralobar and extralobar pulmonary sequestrations. Intralobar sequestrations are located within the same investing visceral pleura as the adjacent normal lung tissue (Fig. 6). Extralobar sequestrations have their own visceral pleura, which physically separate them from the nearby normal tissue. Incidence is 0.15\% to 6.4\%.\textsuperscript{138} The differential diagnosis for this pulmonary malformation includes CCAM, bronchogenic cyst, CLE, teratoma, enteric duplication, and neural crest tumors.\textsuperscript{136,139,140}

Embryologically, normal lung development begins at about 4 weeks’ gestation with the emergence of the tracheal bud from the ventral aspect of the primitive foregut. The tracheal bud progresses in a caudal direction and eventually bifurcates to form the primary bronchial buds. The tracheoesophageal septum forms to separate developing lung from the foregut and thus defines the trachea and esophagus as separate structures.\textsuperscript{141} Multiple generations of bifurcation lead to the development of the normal tracheobronchial tree and are intimately associated with similar branching of the developing pulmonary arteries. Pulmonary sequestrations are believed to arise as supernumerary lung buds that develop caudally to the main lungs. Because these extra lung buds are typically not associated with normal pulmonary artery development, they develop a separate systemic blood supply.\textsuperscript{139} If this process occurs before the development of the visceral pleura, then the aberrant pulmonary tissue becomes invested in the same visceral pleura as the normal lung tissue and is termed intralobar. If the process occurs after the development of the pleura, the sequestration develops its own visceral pleura and is termed extralobar. The arterial supply usually arises directly off the aorta and may arise from below the diaphragm in 20\% of cases. The venous drainage displays more variation. Venous drainage may be via the pulmonary veins, vena cava, azygous or hemiazygous veins, or a combination of some or all of these.\textsuperscript{136,139}

Intralobar sequestrations are the most common type (75\%) and are typically located in the medial and posterior left lower lobe.\textsuperscript{138} Intralobar sequestrations have a low rate of associated anomalies. During childhood, these lesions may present with recurrent

![Fig. 6](image_url)

**Fig. 6.** (A) Chest radiograph reveals density in right lower lung field, which was revealed to be an intralobar pulmonary sequestration. (B) CT image shows the intralobar sequestration shown in (A) within the arrows on the patient’s right outlining the lesion. The most medial arrow indicates a systemic artery to the sequestration that arises from the aorta.
respiratory infections as a result of both compression of the adjacent lung tissue as well as inadequate pulmonary toilet through the pores of Kohn. Intralobar lesions have an infection rate reported as high as 91%. Symptoms may progress to lung abscess and hemoptysis if unrecognized. In 10% of intralobar sequestrations, there is a connection to the esophagus, which also predisposes to recurrent infection.

Extralobar sequestrations make up about 25% of all sequestrations. Patients with extralobar sequestrations have a high rate of associated anomalies and therefore are generally diagnosed at an earlier age than patients with intralobar lesions. Up to 40% of patients have some other associated anomaly, including vertebral and chest wall deformities, hindgut duplications, bronchogenic cysts, CCAM, and congenital cardiac malformations. A total of 5% to 15% of patients with congenital diaphragmatic hernia have an associated extralobar sequestration. As indicated by their embryologic origin, up to 15% of these lesions are located within or below the diaphragm, with up to 10% being located within the abdominal cavity. The most common location for an intra-abdominal extralobar sequestration is the left suprarenal area. Air within an extralobar sequestration or a history of feeding intolerance should prompt a search for a connection to the gastrointestinal tract (including the stomach), which may, although it is uncommon, serve as a source of infection.

The overall infection rate for extralobar lesions is reported as low as 14%. Patients may also present with swallowing difficulties, respiratory distress, high-output congestive heart failure, or back pain from torsion.

Prenatal DUS is identifying pulmonary sequestrations with increasing frequency. These lesions typically appear well defined, echogenic, homogeneous, and often with a definable systemic arterial supply. Postnatally, chest radiograph may be normal or show a mass. Sequestrations with cystic components should raise the suspicion of an associated CCAM within the lesion. Air within the lesion may arise from an anomalous connection with foregut structures or from the pores of Kohn, if the lesion is intralobar. MRI and CT are also helpful for establishing the diagnosis and may show a systemic artery not seen on DUS. These imaging modalities are most beneficial for defining anatomy before surgical intervention. Of paramount importance is identifying both arterial and venous drainage from the sequestration before resection. MRI or CT should also be performed in the case of a disappearing lung lesion on DUS. Several reports have indicated that some lesions seen on DUS may seem to regress spontaneously. This situation is believed to be a result of vascular compromise or spontaneous decompression into the adjacent tracheobronchial tree. However, many of these lesions may still be identified postnatally with MRI or CT. A Gastrografin swallow study or esophagogastroduodenoscopy may be helpful in identifying a connection to the esophagus or stomach. Because of significant overlap between sequestrations and other congenital pulmonary lesions on imaging, a definitive diagnosis may not be possible based purely on imaging data. If a suspected sequestration is in the left suprarenal area, it is important to rule out a neural tumor by obtaining urinary catecholamines.

Sequestrations may present in the fetal period with polyhydramnios, hydrops fetalis, mediastinal shift, or hydrothorax. Compression of the esophagus or fetal stomach, the vena cava, or mediastinal shift may lead to polyhydramnios and potential preterm labor. Pulmonary sequestration associated with hydrops is considered potentially lethal. The role for amniocentesis in this setting is unclear. Patients may be considered for fetal intervention if they are between 24 and 30 weeks’ gestation and there is the presence of hydrops. Pulmonary sequestration with associated hydrothorax has a 22% reported survival rate and fetuses showing this should be considered for thoracoamniotic shunting. Postnatally, the degree of associated pulmonary hypoplasia
in the remaining lung tissue and the nature of other congenital anomalies, if present, are the main determinates of outcome.

All symptomatic lesions require surgical excision. Generally, all intralobar lesions and lesions with a cystic component must also be excised. Most investigators suggest that all lesions should be excised by 6 months to 1 year of age both for diagnostic purposes as well as to avoid the risk of infection and malignant degeneration. The latter is a low risk, and is likely a result of the presence of other pathologic tissue in the presence of an intralobar sequestration. Any infection should be adequately treated before excision. Surgical excision requires careful dissection of the vascular stalk, with ligation of the feeding vessels. The aberrant arteries tend to be large, elastic, and thin walled. Exsanguination may occur if a large feeding vessel of infradiaphragmatic origin retracts below the diaphragm before achieving adequate vascular control. Classically, intralobar lesions are removed via thoracotomy and require a segmental lung resection at minimum. Large intralobar lesions may be best served by lobectomy. The short-term loss of normal pulmonary tissue is offset by compensatory lung growth from the remaining lung. Thoracoscopic approaches are also safe and are associated with decreased postoperative pain, faster recovery, and a lower rate of chest wall deformity. Extralobar sequestrations may be removed without any damage to the adjacent lung. Diaphragmatic or intra-abdominal sequestrations may require laparoscopy or laparotomy for adequate excision. In the rare case of bilateral sequestrations, bilateral excision may be performed during the same procedure, sometimes from 1 side, or as a staged procedure. Embolization procedures have also been described via both the femoral and umbilical route. This approach may be best used for patients who are not stable enough to undergo resection, such as those in high-output heart failure.

**CLE**

CLE is a condition characterized by hyperinflation of otherwise normal alveoli. Most cases are discovered either prenatally, or within the first 6 months of life. Traditionally, this condition has been most commonly recognized in White men, with a male/female ratio ranging from 2:1 to 3:2 reported. This finding has supported a theory that there may be a genetic cause for this condition. However, a series of 21 non-White Arab children with this condition reported in 2001 has brought this theory into question.

The left upper lobe is the most common location of CLE. The right upper and middle lobes are the next most commonly involved. The lower lobes are not commonly affected. This latter point may help in differentiating CLE from CCAM when there is an abnormal degree of lucency in a lower lung field. Causes of CLE are believed to be both intrinsic and extrinsic. Intrinsic causes include bronchial cartilage dysplasia, deficiency, or atresia, bronchial mucosal proliferation, bronchial torsion, and granulation tissue or inspissated meconium or mucus within a bronchus. External causes include compression from aberrant vascular structures, hilar lymph nodes, or mediastinal cysts. A condition known as polyalveolar lobe, in which there is a proliferation of expanded alveoli within a lung lobe, may be a cause of some cases of CLE. About 50% of children have no identifiable inciting cause.

CLE can be suspected by the appearance of cystic changes in a fetal lung on prenatal ultrasonography. However, a diagnosis of CLE is not possible because abnormal cystic lesions seen on such studies may also represent CCAM, lung sequestration, or congenital diaphragmatic hernia. Cystic changes seen prenatally may appear to diminish in size or even disappear. However, postnatal imaging is imperative.
to further assess the nature of the lesions seen. It cannot be presumed that they have resolved.\textsuperscript{137,148,153}

Presenting symptoms and signs of CLE may include dyspnea, tachypnea, wheezing, cyanosis, and tachycardia. Symptoms are not commonly present at birth, and typically progress gradually. However, they can progress rapidly in some instances, particularly in infants. Rapid progression of symptoms is not common in older children.\textsuperscript{144}

A chest radiograph shows a hyperlucency in the area of the involved lung lobe. The presence of some lung markings helps to differentiate this finding from that of tension pneumothorax. There is often mediastinal and tracheal shift and herniation of the lung across the midline. CT or MRI can further delineate the involved anatomy and also reveal obstructing lesions of the involved bronchus such as enlarged lymph nodes, a mediastinal cyst, or an aberrant vascular structure (Fig. 7). Once CLE is believed to be present, bronchoscopy may also be useful in determining the cause of the CLE. Flexible bronchoscopy may be better than rigid bronchoscopy in a controlled setting so as to minimize the use of positive pressure. Identification of abnormal bronchial anatomy may reinforce the diagnosis of CLE as opposed to an intrinsic parenchymal disease. In addition, if inspissated meconium, mucus, or granulation tissue is found, the potential exists to relieve the obstruction and resolve the CLE. Evaluation of CLE should also include an investigation for any congenital heart anomalies because they present in 10\% to 20\% of children with CLE.\textsuperscript{144,146}

It is important not to confuse CLE with tension pneumothorax in a symptomatic patient, because placement of a chest tube into an emphysematous lobe may exacerbate the symptoms. In addition, if intubation is required, positive pressure should be kept at a minimal level so as to minimize further expansion of the involved lobe and avoid worsening pulmonary compromise. Management of CLE has most commonly involved removal of the involved lobe. Although this goal has been accomplished successfully using thoracoscopic technique, other investigators have favored an open technique because of the overexpanded lobe and the difficulty this presents with visualization during thoracoscopic technique. Removal of a lung lobe can be accomplished safely in any age group and is tolerated well.\textsuperscript{137,144,154,155} Some

\begin{figure}[h]
\centering
\includegraphics[width=\textwidth]{fig7.png}
\caption{(A) Hyperexpanded right lung shows lung markings throughout, indicating CLE in this infant. The mediastinum is shifted to the left. (B) CT image of the CLE (arrows) shown in the radiograph in (A); this lesion involved the right upper and middle lobes.}
\end{figure}
investigators have advocated observation of patients who are asymptomatic. This practice has been successful in the short-term in a few series. However, others have suggested that lung lobes that seem to harbor CLE may contain other disease, such as CCAM, that places the patient at risk. There have been reports of patients having lobectomies for a diagnosis of symptomatic CLE only to have other pathologic tissue, such as that indicating CCAM, found within the specimen. Furthermore, if lung lobe removal is needed beyond the infant and toddler years, a window for compensatory lung growth may be lost. Long-term follow-up involves repeated radiologic studies that may present potential risk. In general, long-term results of nonoperative management beyond childhood are not known.

Esophageal Atresia and Tracheoesophageal Fistula

Esophageal atresia and tracheoesophageal fistula (EA-TEF) refer to a constellation of anomalies that result from improper embryogenesis of the foregut. Historically, these lesions were believed to be caused by inappropriate fusion of invaginating lateral longitudinal ridges, which created a septum dividing the foregut into the trachea and the esophagus. However, more modern evidence suggests that foregut development is more intimately involved with the respiratory tract development itself. Adriamycin-induced EA-TEF in rats suggests that the fistula and lower esophagus is of respiratory origin. Another theory holds that abnormalities in signaling of the organ differentiation-promoting glycoprotein, sonic hedgehog, play a role in these esophageal anomalies. Therefore, further understanding is needed about the origin of these anomalies.

EA-TEFs affect males and females equally and occur in about 1 per 3000 to 5000 live births. Infants affected by EA-TEF are premature in 35% of cases. A variety of anatomic areas are known to have a high incidence of anomalies in infants born with EA-TEF. This constellation was originally given the acronym VATER association. This term was later expanded to include the recognized increased incidence of congenital cardiac anomalies and therefore was denoted VACTERL: V, vertebral; A, anorectal; C, cardiac; TE, tracheoesophageal; R, renal; L, limb; lumbar. EA-TEF may also be associated with a variety of other conditions, including Down syndrome, DiGeorge sequence, Fanconi syndrome and the CHARGE association (coloboma, heart defects, atresia choanae, developmental retardation, genital hypoplasia, ear deformities).

The presence of EA-TEF, when not an isolated fistula, may occasionally be detected on prenatal ultrasonography. Findings of a small stomach, polyhydramnios, or upper pouch dilation may be evident. Fetal MRI may be a useful tool in showing EA-TEF prenatally when suspected. Delivery method is based on the obstetric needs of the mother and baby, but delivery at a tertiary care center is typically desired.

EA-TEFs are typically classified into 5 basic types, although many variations may exist. The 5 types and their relative incidences are shown in Fig. 8. Type C is the most common. Type D may occur more frequently than originally believed because many of what are believed to be recurrent TEFs after repair of type C EA-TEF are missed proximal fistulae of type D.

Symptoms of EA-TEF are usually evident at birth. If esophageal atresia is present, the infant often shows excessive drooling at birth. Any attempt at feeding results in regurgitation and choking. Aspiration may result in cyanosis and respiratory distress. An attempt to pass an orogastric tube results in resistance. If a distal fistula is present, the abdomen may become distended because of inspired air filling the stomach through the fistula. This situation may be exacerbated by any positive pressure generated by hand-mask ventilation, or positive pressure ventilation through an
endotracheal tube. In addition, pulmonary compromise may result from reflux of gastric contents into the airway through a distal fistula, or from spill-over of saliva from the proximal atretic pouch. A chest radiograph that includes the neck shows the end of a tube placed through the mouth or nose into the esophagus to be at the level of the lower neck or proximal thorax. Instillation of a small amount of air can help outline the proximal atretic esophageal pouch. An abdominal radiograph shows the presence of air in the intestine if there is a distal TEF. However, the absence of air typically indicates the presence of isolated esophageal atresia. This finding is the same in the rare instance of esophageal atresia with only a proximal fistula. The length of the proximal pouch, and the presence or absence of a proximal fistula, can be determined by instillation of 0.5 to 1 mL of thin barium under direct fluoroscopic vision (Fig. 9). A proximal fistula can also be assessed for using bronchoscopy at the time of surgery if necessary.

Isolated TEF without EA is usually a delayed diagnosis because the esophagus is intact, and babies often tolerate early feeds. However, these babies tend to have frequent coughing with feeding and are prone to recurrent respiratory infections. The diagnosis can be established by a contrast esophagogram. However, it is important to specifically discuss with the radiologist that an isolated TEF is being sought because the baby must be prone, and possibly in a small amount of Trendelenburg position, to define this fistula with the contrast. This radiographic strategy is used because the fistula typically courses from the esophagus in a cephalad direction to the trachea. Bronchoscopy or esophagoscopy may be required if suspicion remains high and the contrast study is negative.

When EA or TEF is established, further workup is required to assess for any of the anomalies of the VACTERL association. Evaluation should include a thorough physical examination, looking particularly for any anorectal anomalies, a cardiac echo, a renal echo, chromosomal studies, and appropriate plain radiographs to assess the vertebral column and limbs as needed.

In the preoperative period, the infant should be positioned in a semiupright position. A suction catheter should be placed in the atretic esophageal pouch, and intravenous fluids and antibiotic coverage should be provided. It is preferable to avoid intubation if

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**Fig. 8.** Classification of esophageal atresia or tracheoesophageal fistula (from left to right): type C, esophageal atresia with distal tracheoesophageal fistula (85%); type A, esophageal atresia without fistula (8%); type B, esophageal atresia with proximal fistula (1%); type D, esophageal atresia with proximal and distal fistulae (2%); type E, tracheoesophageal fistula (H-type) without atresia (4%). (Adapted from Bax K. Esophageal atresia and tracheoesophageal malformations. In: Holcomb III GW, Murphy JP, editors. Ashcraft’s pediatric surgery. 5th edition. Philadelphia: Saunders; 2010. p. 346; with permission.)
possible, but if intubation is required for airway support, it is desirable to keep the positive pressure as low as possible, and even allow the baby to breathe on its own if possible. In the rare instance that an infant’s respiratory status is unduly compromised by abdominal distention, an emergent gastrostomy tube can be placed for decompression.

Operative intervention can usually be undertaken in an elective or semielective manner. The particular operation is determined by the lesion present. Type C and type D anomalies can usually be repaired primarily. The traditional approach has been via a right-sided thoracotomy through the fourth intercostal space. A muscle-splitting incision may preserve most or all of the latissimus dorsi and serratus anterior muscles and reduce the incidence of muscle atrophy and scoliosis. A retropleural dissection facilitates exposure. The fistula (or fistulae) is divided and a primary anastomosis of the esophagus is accomplished with a single layer of suture. This repair has also been accomplished via a thoracoscopic technique.  

The repair can be assessed at 5 to 7 days postoperatively with an esophagogram. If there is no leak, and the anastomosis is acceptably patent, feeding by mouth can begin and any chest drainage tubes can be removed.

Occasionally, a right-sided aortic arch may be discovered on preoperative cardiac echo, or at operation. Repair can be undertaken either through a right-sided or left-sided approach. However, these infants are prone to having a long-gap atresia, double aortic arch, or other arch anomalies. If a long-gap atresia is found at operation for a type C or D lesion, it is best to divide the fistula(e), attach the lower esophagus to the prevertebral fascia, and place a gastrostomy feeding tube. The infant can then be supported with a suction catheter in the proximal pouch and gastrostomy feeds for the next 8 to 12 weeks. The 2 ends of the esophagus typically grow closer together over this time, or at least grow stronger, thereby optimizing the opportunity to use the native esophagus at final repair. Some investigators advocate techniques of bougienage of the proximal pouch and bolus feeding of the stomach to stretch the esophagus at both ends as much as possible. Techniques at operation for maximizing esophageal length include mobilizing both the proximal and distal

Fig. 9. The atretic proximal pouch of esophageal atresia is outlined by instillation of thin barium (arrows) under direct fluoroscopy. No proximal fistula is found.
esophageal limbs, and circular or spiral myotomy for the proximal limb. External traction on the proximal and distal limbs has also been reported. If the esophagus cannot be approximated, operative strategies can include gastric pull-up, gastric tube formation, and colon interposition.171–178

Operative strategy for isolated esophageal atresia, or EA with proximal TEF (types A and B), is the same as that for long-gap atresia found with type C or D, as described earlier. Types A and B commonly have a long gap between the 2 ends of the esophagus. If a proximal fistula is present, bronchoscopic and esophagoscopy evaluation can localize it, and the fistula may be divided through the neck.

Isolated TEF can usually be repaired through a neck approach. Bronchoscopy is used to place a ureteral catheter through the fistula into the esophagus. The infant is then intubated, and the catheter is retrieved via esophagoscopy. The catheter is then used to localize the fistula by both traction and fluoroscopic examination. Care must be taken to preserve the recurrent laryngeal nerves during the division of an isolated TEF and the subsequent repairs of the esophagus and trachea. Surgical division may be accomplished via a thoracoscopic technique.179 Obliteration of an isolated TEF using a yttrium aluminum garnet laser or electrocautery has also been described.180

Prognosis for outcomes from EA–TEF repairs has been classified in many ways, and with modifications, over the years. The Waterston and Spitz classifications have based prognosis primarily on birth weight and the presence or absence of cardiac disease. Overall, survival has improved markedly over the years, and in general, with modern intensive care unit support and surgical technique and strategies, outcomes are favorable, rendering these classifications potentially obsolete.158

Complications of repair of EA or TEF include (with reported ranges) leak (5%–20%), stricture (6%–40%), dysphagia (50%–75%), reflux (30%–60%), recurrent fistula (10%), chest wall deformity (5%), and scoliosis (5%).181,182 Esophageal motility is not normal in patients with esophageal atresia, and parents must be counseled to provide food that infants and children can chew and swallow well. Strictures can usually be balloon-dilated under fluoroscopic guidance. The advantage of this strategy over bougienage is that the dilation is performed under direct vision with controlled pressures, and any leak can be studied immediately.183,184 Recurrent fistulae have traditionally been repaired via thoracotomy, sometimes using a pericardial, pleural, or azygous vein flap. However, recurrent fistula rates after thoracotomy for initial recurrence approach 20%.185–187 Successful closure of a recurrent fistula using a bronchoscopic technique has been described.188

Increasing information is being gathered regarding the long-term outcomes of EA–TEF repairs because some of the patients repaired in the early days of these repairs are now adults. Many of these patients have symptoms of gastroesophageal reflux, dysphagia, and occasional compromised passage of food. They have learned to accommodate this in many instances. In addition, endoscopic studies have shown that some patients do have inflammatory or metaplastic changes in their esophageal mucosa. However, there are no reports linking any increase in esophageal cancer to patients having undergone EA–TEF repair.179,189–191 In addition, when symptoms have been recognized in these patients, they seem to dissipate noticeably in many patients over the long-term.181,182

REFERENCES


