The Esophagus

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2.1 Anatomy and Development of the Esophagus

2.1.1 Development

The esophagus and trachea form from a single common tubular structure that separates into an anterior laryngotracheal groove and a posterior esophagus (Collins 1995). Development of the esophagus begins in the fourth week of fetal life when the respiratory diverticula, or lung buds, appear on the ventral wall of the foregut at the border with the pharyngeal gut. Lateral invagination of the mesodermal esophagotracheal septum gradually partitions this diverticulum from the foregut, thus dividing into the ventral respiratory primordium and the dorsal esophagus. Epithelial growth obliterates the lumen, which recanalizes at the tenth week (Sadler 2000). The esophagus is initially short, but lengthens rapidly with descent of the heart and lungs, and attains normal length by 7 weeks. Occasionally, the esophagus fails to lengthen sufficiently, and the stomach is pulled up into the esophageal hiatus through the diaphragm, resulting in a congenital hiatal hernia (Sadler 2000). The commonest congenital anomalies, i.e. the tracheoesophageal fistula/esophageal atresia complex, esophageal stenosis, duplication cysts and bronchopulmonary foregut malformations, are the result of disruption of the normal sequence of separation of the airway from the foregut. The upper esophageal sphincter and normal peristaltic activity are well developed by 33 weeks, and continue to mature in the post-natal period (Jadcherla et al. 2005).

2.1.2 Anatomy

The esophagus is a muscular tube that connects the pharynx at the level of the inferior border of the cricoid cartilage at C6 to the cardia of the stomach at the level of T11 (Meschan 1975). The course of the esophagus is approximately vertical, with a gentle curve to the level of the lower neck. The esophagus gradually returns medially at the level of T5, inclining again to the left at the level of T7, and finally inclining anteriorly to traverse the diaphragm at T10 (Bannister 1995). The esophagus lies anterior to the spine, and its course parallels the curves of the cervical and thoracic spine. Apart from the appendix, the esophagus has the narrowest lumen of the alimentary tract. Four anatomic sites of constriction occur at which the esophageal lumen becomes even narrower. These include: at the cricopharyngeus muscle, the level where the aortic arch crosses the esophagus, the level at which the left main stem bronchus crosses the esophagus just below the carina, and the level of the esophageal hiatus in the diaphragm. A foreign body or a large food bolus may lodge at one of these natural constrictions and thereby obstruct the esophagus. The lower esophageal sphincter, or vestibule, is at the level of, or below, the diaphragm and is held in place by the phrenico-esophageal membrane. The lower esophageal sphincter is formed by smooth muscle fibers that surround the intra-abdominal esophagus. These fibers remain contracted except during swallowing and vomiting. Sphincteric action is also provided by the diaphragmatic crura that surround the esophagus. The crural muscle fibers constrict the esophagus during inspiration and when intra-abdominal pressure is raised and prevent gastroesophageal reflux (Bannister 1995). The acuteness of the angle of the gastroesophageal junction also prevents reflux.

The esophagus is lined by a thick mucosa of non-keratinized squamous epithelium that provides an impermeable barrier and, together with surface mucus, protects against mechanical injury. The muscularis mucosae is made up of longitudinal smooth muscle that becomes thicker distal to the pharynx. The mucosa is loosely attached to the muscularis mucosae. The muscularis consists of an outer, thicker longitudinal layer and an inner circular layer. Accessory slips of muscle fibers attach and anchor the esophagus to adjacent thoracic organs and to the pleura. Striated skeletal muscle occurs in the upper two-thirds of the esophagus, while only smooth muscle is found distally. In the empty state, the mucosa is thrown into a series of parallel longitudinal folds. It is these folds that give the esophageal lumen a stellate appearance so characteristic in cross section. Mucosal folds are not a feature of the neonatal esophagus, which is smooth in outline.

2.2 Radiological Techniques of Examination

An empty esophagus is not visible on plain radiographs or CT. However, the esophagus is not uncommonly outlined by air in a child that is crying and swallowing large amounts of air (Fig. 2.1). An air-filled esophagus is also frequently seen in neonates ventilated with continuous positive airway pressure (CPAP), as well as in those with tracheo-esophageal fistula, esophageal stricture and achalasia (Fig. 2.2). In children with developmental delay, air in the esophagus is a common finding, and is usually secondary to reflux.

Despite the current emphasis on the superb diagnostic possibilities of CT and MR, the conventional barium examination still remains the most important modality for evaluating patients with dysphagia, gastroesophageal reflux and other symptoms referable to the esophagus (Levine and Rubesin 2005). The barium swallow, or esophagram, provides anatomic and functional information about the esophagus along its entire course. Radiation safety and dose reduction are important factors to consider for pediatric imaging, and fluoroscopy time must be carefully monitored. Dose reduction is achieved with low-dose pulsed fluoroscopy. "Last image hold" on the monitor helps to reduce fluoroscopy exposure. Because fine mucosal detail is not the main object in routine esophagrams, image capture from the disc provides sufficient information without additional radiation.

Barium suspension is the most frequently used contrast medium. Children above 6 months of age who are reluctant to drink barium may be encouraged by adding additional flavoring to the barium. Any commercially available chocolate syrup or instant drinking chocolate powder renders the barium more palatable without altering its radiographic characteristics. It is advisable to check for allergies and to be aware that some commercial flavoring preparations contain allergenic products.

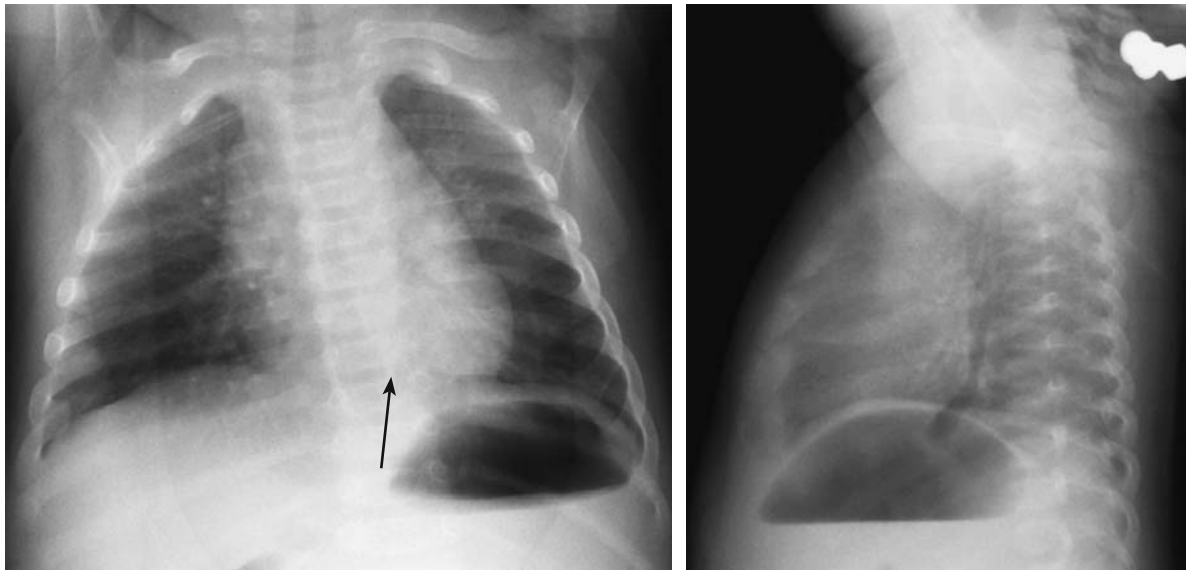


Fig. 2.1a,b. Air outlining a normal esophagus. AP (**a**) and lateral chest (**b**) radiographs of a crying infant with an air-filled esophagus (*arrow*)

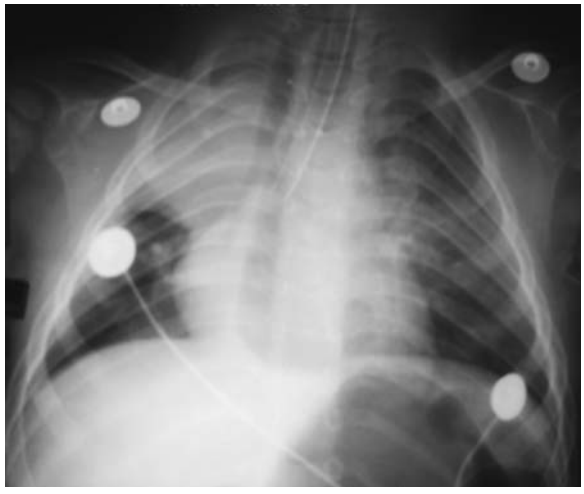


Fig. 2.2. Air-filled esophagus. Esophageal malposition of the endotracheal tube with resultant gaseous gastric and esophageal distention and right upper lobe atelectasis

Some children find chilled barium more palatable. Barium must not be used if either esophageal perforation or massive aspiration is suspected. Such patients are best studied with water-soluble low osmolar non-ionic contrast media. Gastrografin, due to its high osmolality, should never be used in infants or in any patients lacking adequate airway protection. Its use must be restricted to older, stable and neurologically unimpaired patients.

Children who are to undergo barium swallow examinations must be fasting. A child who is not hungry will simply refuse to drink barium. The duration of fasting depends on the child's age, and should be no longer than the child's routine time between feeds. Premature infants should fast 2–3 h, and infants up to 3 months fast 3–4 h. Children above 2 years can fast up to 6 h. To minimize parental and patient discomfort, fasting children are best scheduled for examinations early in the morning. Older children and adolescents will be most comfortable swallowing barium in the erect position. Infants and younger children are examined recumbent, and require immobilization. Effective immobilization decreases fluoroscopy time, and ensures clear and diagnostic images. Safe and convenient immobilization is accomplished with a device such as the Octagon board (Octostop, Laval, Qc, Canada) which enables immobilization, as well as rotation of the child into any position, including true lateral and oblique. This device facilitates positioning with the infant's arms above the neck so that the esophagus is not obscured during fluoroscopy. Barium can be very conveniently administered with the modified Poznanski technique using an 8-F feeding tube inserted through the end hole of a nipple and injected via syringe (Kuhns and Poznanski 1972; Poznanski 1969). The side holes of the feeding tube are further from the tip, so the tube must be advanced all the way through the nipple and

the protruding 1-cm cut so that the tube is almost flush with the nipple in order to prevent gagging. When the nipple is loosely attached to the face with paper tape, this "hands free" technique ensures that no hand or bottle obscures the mouth or pharynx during the examination and also decreases exposure to the radiologist. Barium is injected at a rate that is easily judged by observing a few trial sips of contrast medium prior to commencing fluoroscopy. This rate of administration of barium should be maintained to prevent aspiration. Older children may be fed from a cup or they may drink through a straw if they are examined in the upright position. When evaluating for gastroesophageal reflux, the volume of barium administered should be the same as that of a normal feed. If the child refuses to drink sufficient barium, the volume can be augmented with milk or juice after the anatomy to the level of the duodenojejunal flexure has been evaluated.

Swallowing begins in the mouth when chewed food or liquid reach the back of the tongue which then elevates and propels the oral contents posteriorly to the pharynx while the soft palate elevates to occlude the nasopharynx. The larynx and hyoid bone are seen to elevate as the oropharynx contracts, propelling the bolus distally while the epiglottis closes to protect the airway. The oral and pharyngeal phases of swallowing are evaluated in the lateral position to avoid overlap with the skull and spine. Peristalsis begins in the cervical esophagus, and the bolus passes inferiorly with a smooth primary stripping wave (Schlesinger and Parker 2004). Distal to the cervical esophagus, the esophagus is evaluated in the oblique position and true lateral, thus projecting the esophagus off the spine and avoiding overlap. Gravity helps the esophagus to clear, and infants with poor esophageal motility may benefit by tilting the fluoroscopy table into a more upright position. The distended esophagus has a smooth and regular outline. In its collapsed state, the mucosal folds appear as parallel, longitudinal lines 1–2 mm thick (Fig. 2.3). The esophagram in children should always include the stomach and the duodenum to the level of the duodenojejunal flexure to observe gastric emptying and to document normal bowel rotation. Cross-sectional imaging has limited application in evaluation of the esophagus, but is invaluable in cases with extrinsic compressive masses and vascular rings.

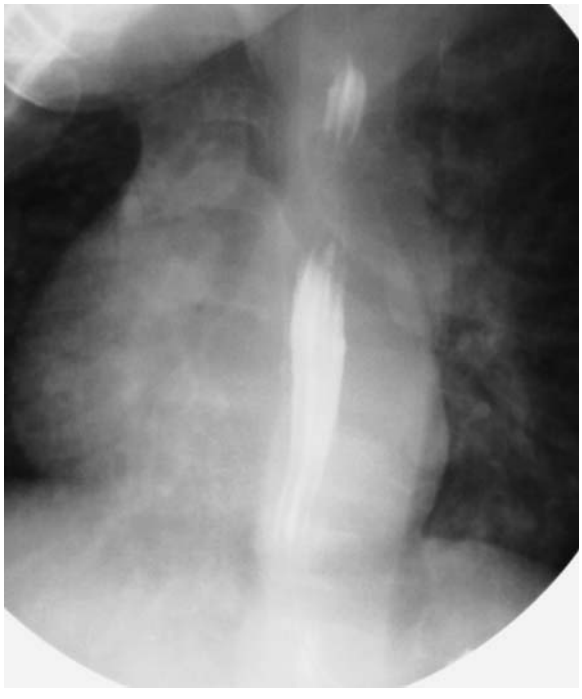


Fig. 2.3. Normal mucosal folds. Parallel linear mucosal in an infant's empty esophagus

2.3 Disorders of Swallowing

Feeding difficulty is not an uncommon symptom in children, reportedly occurring in 25% of children (Miller and Willging 2003). The causes of pediatric dysphagia are varied and complex, and may be physiologic or behavioral. The majority of children with dysphagia have a neurological cause, and may be due to cranial nerve palsies, cerebral palsy and meningomyelocele. Structural craniofacial anomalies predispose to dysphagia (Lifschitz 2001; Miller and Willging 2003). Swallowing dysfunction with aspiration is common in full-term infants less than 1 month of age and improves with age (Vazquez and Buonomo 1999).

The video esophagram, or modified barium swallow, is the standard technique to evaluate dysphagia. This examination is more sensitive than clinical evaluation of aspiration (DeMatteo et al. 2005), and is also more sensitive than the conventional upper GI series for the detection of aspiration (Vazquez and Buonomo 1999). A scout radiograph of the chest should be obtained to assess for evidence of aspiration. For the examination, the child must be securely

maintained sitting in the true lateral position, best accomplished in an infant feeding seat. Participation of the parents and caregivers helps to reassure the child, and recreates some aspect of daily feeding. The child's own speech therapist or feeding therapist should optimally be present during the examination to witness the events. The therapist can observe the optimal food volume and consistency and compensatory maneuvers that assist swallowing (Fernbach 1994). Boluses of different consistencies are fed. Young children are given liquids, though the density of various liquids may vary. The examination in older children begins with thin liquid barium, proceeding to feeds with a mixture of barium thickened with pudding or pureed food, and finally with more solid food such as barium-coated crackers. Barium density influences the swallowing mechanism. High-density barium has a slower transit time, causing the upper esophageal sphincter to open later, to remain open for longer and to delay its closure (Dantas et al. 1989). The actions of swallowing occur too rapidly to be observed fluoroscopically, therefore the entire examination is recorded on videotape, or if unavailable, with standard fluoroscopy set at a frame rate of 2–3/s. The recorded examination can be repeatedly reviewed without additional radiation. Review of the tapes is important when there is a need to assess the patient multiple times as treatment or the disease progresses. Multiple swallows in each series must be reviewed because changes can occur in the same cycle after episodes of normal swallowing. Infants may tire as feeding progresses, and may have difficulty maintaining their airway so the study should be continued after the first few uneventful swallows (Newman et al. 2001). Pulsed fluoroscopy cannot be used during video esophagrams as it may prevent detection of fleeting episodes of penetration and microaspiration (Mercado-Deane et al. 2001). The unfortunate necessity of using standard fluoroscopy results in the child's receiving a higher radiation exposure to the thyroid gland than during a conventional esophagram.

Abnormalities occur at all levels during swallowing: in the oral, pharyngeal and esophageal phases. The modified barium swallow is used to identify the level of pathology so that therapy or treatment can be given. In the oral phase of swallowing, the abnormality may have an anatomic etiology such as micrognathia or macroglossia. Children on long term tube feeding may be unused to feeding, and they may simply refuse to eat. Severely neurologically impaired children may be unable to suck or they may lack sufficient tongue control to latch onto and maintain control of the nipple. Suckling deficits also manifest with weak and deficient tongue motion, inability to compress the nipple and early tiring (Kramer 1989). Oral motor dysfunction occurs with moderate or severe cerebral palsy and developmental delay. Incomplete buccal closure leads to drooling, and abnormal tongue and jaw motion (Kramer 1989). Those neurologically impaired children unable to elevate the soft palate experience nasopharyngeal incoordination and nasopharyngeal reflux (Fig. 2.4). Occasional nasopharyngeal incoordination is most commonly due to transient swallowing incoordination. Retropharyngeal masses such as teratoma, lymphoma and abscess can rarely cause dysphagia. Cricopharyngeal achalasia, or failure of relaxation of the cricopharyngeus muscle, is most commonly secondary to gastroesophageal reflux. Signs of cricopharyngeal achalasia include absent, delayed or incomplete opening or early closure of the sphincter. Laryngeal penetration occurs when barium enters an incompletely protected airway below the level of the vocal cords and trachea during swallowing. It is important to document whether aspiration induces

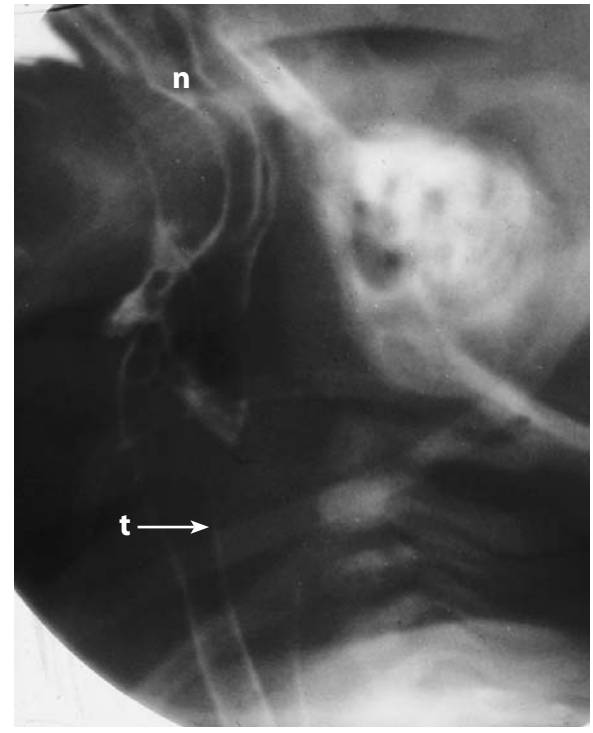


Fig. 2.4. Nasopharyngeal and tracheal aspiration. Barium outlines the nasopharynx (*n*) and the trachea (*t*). Only a trace of barium remains in the esophagus

a cough reflex. Penetration is different from aspiration, which occurs during respiration (Kramer 1989). Aspiration should be documented with a post-examination chest radiograph. The examination must be terminated if aspiration occurs and causes changes in vital signs. If the child remains stable after aspiration, the examination can proceed. The aim of the study is to determine if there is a safe way that food can be given, and after aspiration, it may be necessary to change to barium of another density, to use another type of nipple or to change the pace of feeding. Co-ordination of swallowing improves with age, and follow up examinations are an effective means to monitor improvement.

2.4 Congenital Abnormalities

2.4.1 Esophageal Atresia and Tracheo-esophageal Fistula

Esophageal atresia (EA) and tracheo-esophageal fistula (TEF) are common congenital anomalies, encountered in approximately 1 in 3,500 births (Shaw-Smith 2005). They are part of a spectrum of congenital anomalies that arise from defective formation of the esophagus or abnormal communication between the esophagus and trachea. It has been suggested that tracheo-esophageal fistula is the result of incomplete infolding of the lateral mesodermal walls that separate the developing esophagus from the trachea. Atresia is thought to result when the lateral folds of mesoderm occlude the esophageal lumen (Berrocal et al. 1999). Intrauterine anoxia and vascular compromise can cause focal esophageal necrosis that may result in atresia or a fistulous connection between the esophagus and adjacent trachea (Barnard 1956). Five types of anomalies occur: EA with a distal fistula to the trachea, accounts for more than 80% of cases, esophageal atresia without a fistula occurs in 10%, and an isolated H-fistula accounts for 6%. Atresia with a distal and a proximal fistula occurs in about 1%, and the rarest, atresia with a proximal fistula, accounts for fewer than 1% (Cumming 1975). Children with esophageal atresia have the most urgent clinical presentation due to their inability to swallow salivary secretions which then overflow and are aspirated into the airway. Respiratory distress may be exacerbated by gastroesophageal reflux from the distal esophageal segment into the airway when a distal fistula is present. Inability to pass an orogastric tube confirms the clinical diagnosis. Fistulae between the esophagus and trachea are in the midline. Although classically described as an H-fistula, the connection to the esophagus has an upward, oblique course that assumes an N-shape. A small isolated fistula without atresia may be less symptomatic and asymptomatic until late childhood (Kappelman et al. 1969) whereas a large fistula will be more likely to present earlier. Intrauterine distention of the blind-ending esophageal pouch compresses the adjacent trachea, inhibits development of the normal C-shaped tracheal rings and causes tracheomalacia which most commonly affects the proximal two-thirds of the airway. The trachea retains a U-shape with a wide posterior membranous portion, leading to tracheomalacia (Kovesi and Rubin 2004).

EA and TEF are frequently not isolated and occur in association with other congenital anomalies in 25% of children; in children with isolated EA, the incidence of associated anomalies is even higher, affecting 50%–70% of patients (Kovesi and Rubin 2004). The VACTERL association is an acronym for a complex of anomalies that may affect various systems (vertebral, anorectal, cardiovascular, tracheoesophageal, renal and the limbs) (Quan and Smith 1973; Fernbach and Glass 1988). The most frequently encountered anomalies associated with EA and TEF are cardiac (35%), genitourinary (24%), gastrointestinal (24%), skeletal (13%) and central nervous system (10%) (Kovesi and Rubin 2004). Approximately 8% of infants with esophageal atresia have aortic arch anomalies, and approximately 5% of infants with EA have a right aortic arch (Babu et al. 2000). Once the diagnosis of EA is established, prompt evaluation of clinically significant abnormalities (cardiac and renal) should be undertaken. Limb anomalies can be evaluated later as they are not of critical importance.

Until the twentieth century, esophageal atresia was a condition incompatible with life. Survival is now routine, dependent on the severity of other associated anomalies. Treatment of a TEF is surgical closure of the communication. Primary surgical anastomosis is the treatment of choice for EA, but this may be complicated with a long atretic segment. Staged repair is reserved for very ill and unstable children, or those with a long atretic gap (Cumming 1975).

2.4.1.1 Radiological Findings

The radiological appearance varies with the type of lesion, whether there is esophageal atresia, fistula or both. The initial radiograph must include the entire abdomen to assess for the presence of bowel gas (Figs. 2.5 and 2.6). The abdomen is characteristically gasless in the absence of a fistula, whereas in the presence of a distal fistula, the abdomen has a normal bowel gas pattern. Features of esophageal atresia are characteristic. The proximal blind-ending pouch is lucent and distended with air, often with a coiled esophageal tube. The lateral chest radiograph confirms the distended esophagus which displaces the airway anteriorly, markedly narrowing the tracheal lumen (Fig. 2.7). The cervical esophagus can become quite distended in children on nasal continuous positive airway pressure (CPAP) and have a similar appearance to EA (Walor et al. 2005). A right-sided aortic arch occurs in 5% of children with EA/TEF and may be difficult to see on the plain chest radiograph of infants (Berdon et al. 1979). Pre-operative echocardiography should be routinely done in these babies to evaluate the heart and the aortic arch in order to plan surgical access on the side contralateral to the aortic arch. The radiograph should be evaluated for vertebral and limb anomalies (Fig. 2.8).

A "pouchogram" of the atretic proximal esophagus is not usually necessary because of the low incidence a fistula from the proximal pouch (Fig. 2.9). In addition, this is a dangerous procedure if improperly performed. Air or non-ionic isotonic contrast medium can be injected to distend the pouch (Fig. 2.10). Over distention of the pouch with contrast medium will invariably result in aspiration so only a small volume of contrast medium, not exceeding 1–2 ml, is injected into the blind-ending pouch with the child in the true lateral position. Contrast medium must be removed at the end of the procedure.

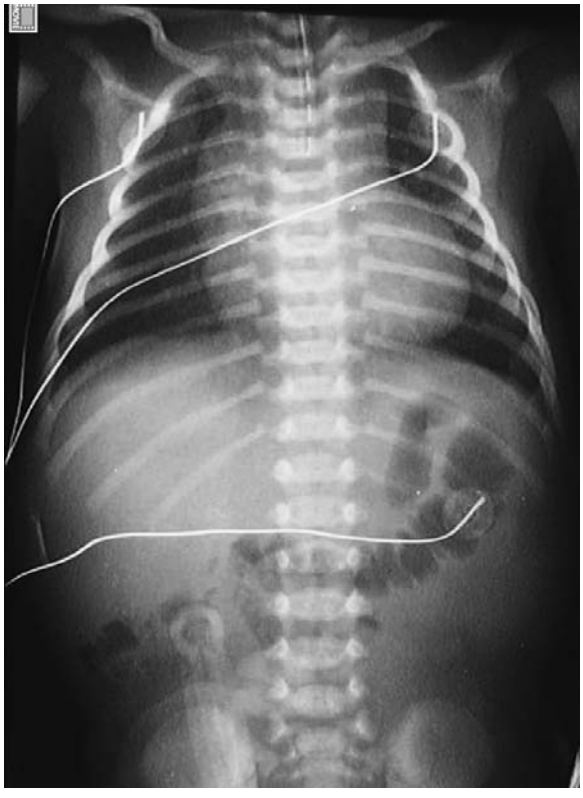


Fig. 2.5. Esophageal atresia with distal tracheoesophageal fistula. The orogastric tube tip is in the blind-ending esophagus and gas is present in loops of bowel in the abdomen. Note the right cervical rib and left 13th rib

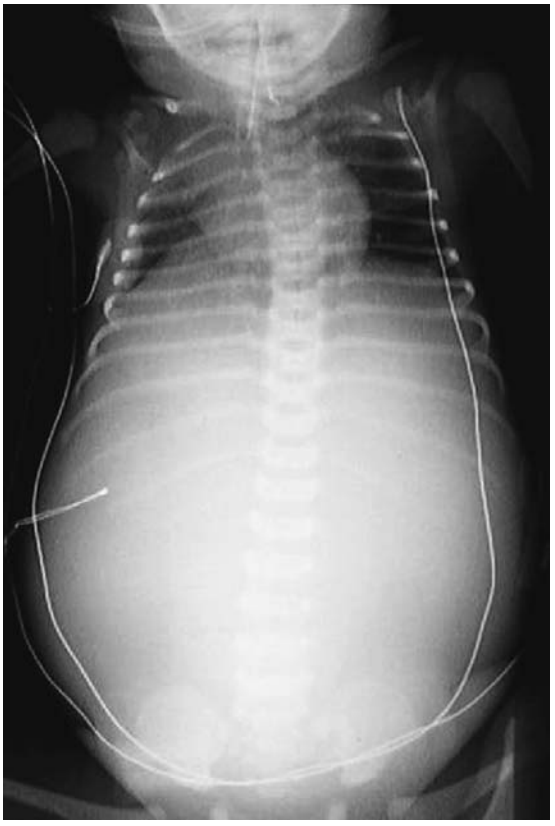


Fig. 2.6. Esophageal atresia without distal tracheoesophageal fistula. The abdomen is gasless. Duodenal atresia was diagnosed at the time of surgical repair

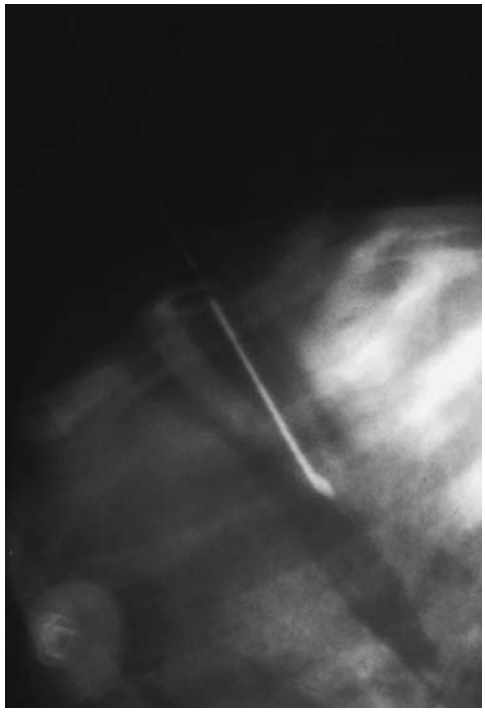


Fig. 2.7. Tracheomalacia. Lateral radiograph with the orogastric tube in the distended atretic proximal esophagus and markedly decreased caliber of the airway at the same level

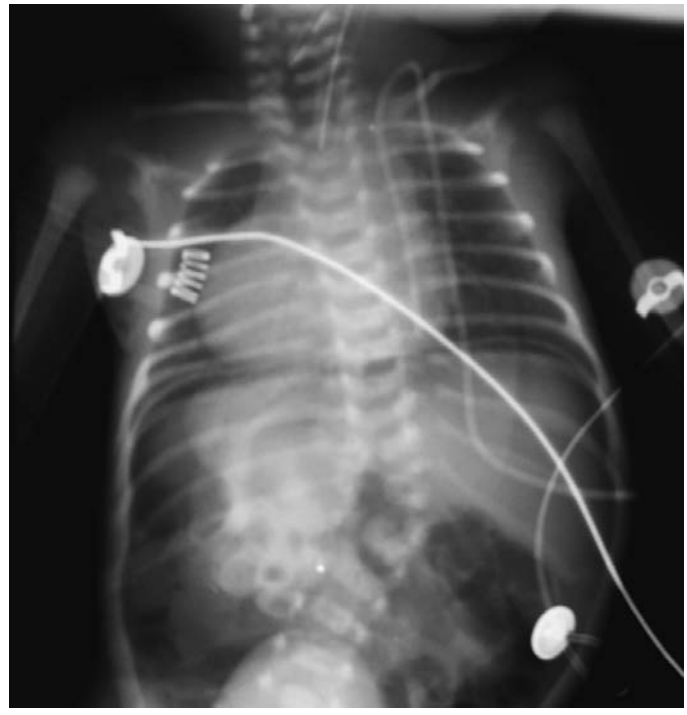


Fig. 2.8. Esophageal atresia with vertebral anomalies. Newborn with EA and a long gap not amenable to primary repair. Pneumoperitoneum followed the recent laparotomy. The esophageal tube is in the blind-ending esophagus. Note the vertebral segmentation anomalies and corresponding missing right sided ribs

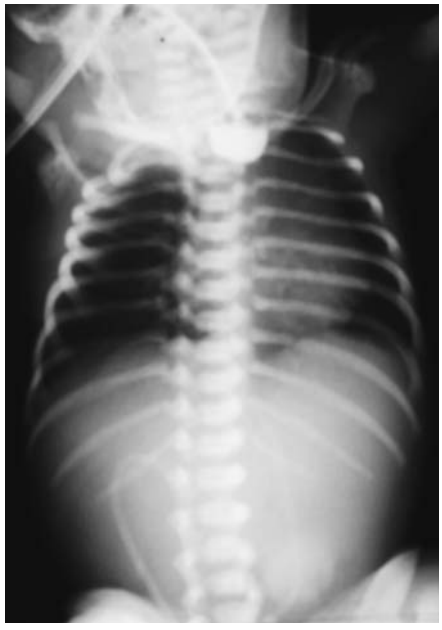


Fig. 2.9. AP "pouchogram". Contrast medium demonstrates esophageal atresia. The abdomen is gasless in the absence of a distal fistula. The left scapula is winged and elevated (Sprengel deformity)

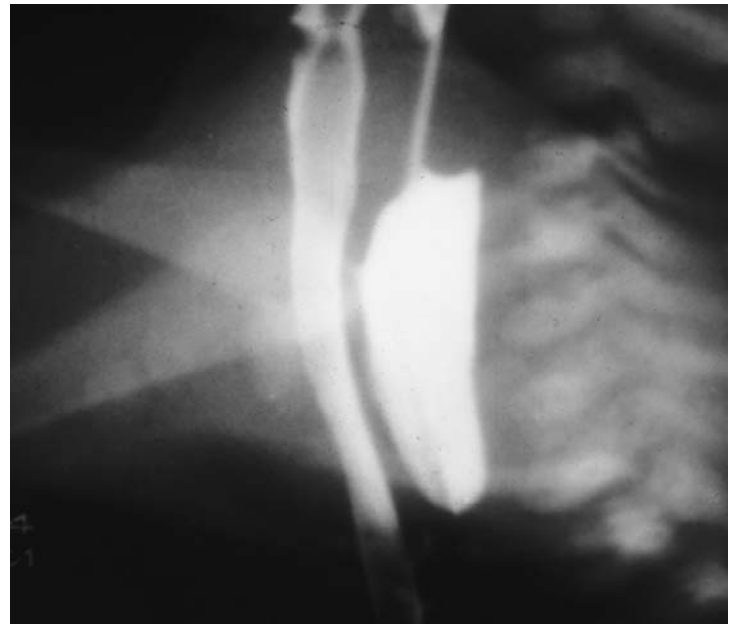


Fig. 2.10. Lateral "pouchogram" with esophageal atresia and proximal tracheoesophageal fistula. Contrast medium fills the blind-ending esophageal pouch and a narrow fistula connects to the trachea

An H-type fistula is often difficult to demonstrate. The barium esophagram, performed to evaluate swallowing and possible aspiration, may reveal the TEF (Fig. 2.11). If the barium esophagram is normal, and suspicion of TEF is high in an infant with recurrent aspiration pneumonias or cyanotic episodes, a more invasive investigation is required. For this procedure, the infant is placed on the octagon board in the true lateral position. Prone positioning is not necessary. The lateral position will give the best unobstructed view of the esophagus and airway, and gives the best access for suctioning. The correct examination technique is with a nasogastric tube inserted into the esophagus to the level of the carina. The tube is slowly withdrawn while low-osmolar non-ionic contrast medium is being injected at a rate sufficient to distend the esophagus. The fluoroscopist must always visualize the larynx during the esophagram to be able to differentiate whether tracheal contrast entered through a fistulous connection or by aspiration. The upper cervical esophagus is the commonest location for an H-type fistula, and it is here that greatest care must be taken in order not to overfill the esophagus and cause spill into the airway and aspiration, and also not to miss a subtle fistulous track. The fistulae are muscular tubes that are not constantly open. They are thought to open during respiration and with swallowing. The fistula will be missed if it is temporarily plugged with food or mucus. It is not uncommon for the fistula not to be demonstrated the first time, thus requiring more than one contrast examination.

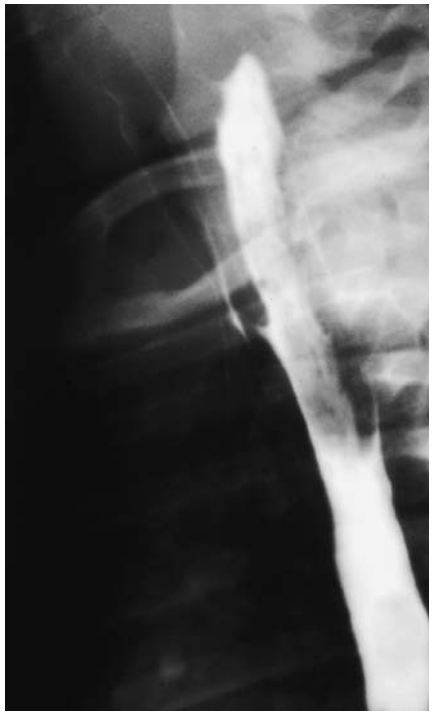


Fig. 2.11. Tracheoesophageal fistula. The fistulous connection to the trachea was opacified during the barium swallow examination. The fistula has an oblique cephalad course

2.4.2 Post-operative Appearance and Complications

The immediate post-operative changes, as well as the later complications, have specific appearances. The underlying abnormalities of the esophagus and airway may produce problems throughout the child's life.

The earliest acute complication of surgery may be an anastomotic leak, occurring in up to 17% of cases (Kovesi and Rubin 2004). Most leaks resolve spontaneously with conservative management: drainage of the leak and esophageal rest. However, up to 50% of those with a prior anastomotic leak will develop an esophageal stricture (Kovesi and Rubin 2004). The commonest late complication in all children with EA and TEF is esophageal stricture, affecting between 6%–40% (Engum et al. 1995). Strictures are more common in the presence of gastroesophageal reflux (GER). Strictures are not uncommonly complicated by foreign body impaction. Strictures, when present, must be differentiated from congenital esophageal stenosis. Re-fistulization occurs in approximately 9% of patients, and may occur as early as 11 days up to 18 months after primary repair (Cumming 1975; Benjamin 1981). Poor esophageal motility and reflux occur in 75%–100% of children with EA and TEF surgery, caused by abnormal in utero development of the esophageal myenteric plexus. GER and dysphagia are common throughout the lives of affected children. GER may be due to the abnormal esophageal myenteric plexus (Kovesi and Rubin 2004) or due to post-operative changes at the gastroesophageal junction. Reflux is exacerbated by poor antegrade esophageal peristalsis. Esophageal obstruction is commonly encountered due to poor esophageal motility or strictures.

2.4.2.1 Radiological Findings

An esophagram with non-ionic water-soluble contrast medium is routinely performed 4–7 days after

surgery to assess the integrity of the anastomosis prior to commencing feeding. The surgical anastomosis is always narrower than the previously obstructed proximal pouch, and should not be confused with a stricture (Fig. 2.12). A normal anastomosis will not impede or obstruct the flow of contrast medium.

A barium esophagram should be performed with a change in eating pattern or new onset of dysphagia that may indicate a stricture. Strictures are usually treated by repeated dilatation, but treatment is less successful when associated with reflux (Kovesi and Rubin 2004). Drooling and refusal to eat are usually signs of foreign body or food impaction. Removal of foreign bodies may be difficult with balloon extraction because of the capaciousness of the esophagus that allows the foreign body to fall back into this segment (Cumming 1975). CT performed years after surgical repair of EA and TEF shows the esophagus to be dilated and filled with air and fluid that is likely a mixture of pooled saliva and refluxed gastric contents, and predisposes to aspiration in the recumbent position (Griscom and Martin 1990). The trachea of these children exhibits more variable cross-sectional variation than in normal subjects (Griscom and Martin 1990). Acquired chest wall deformities and scoliosis are not uncommon after thoracotomy for EA repair (Chetcuti et al. 1989).

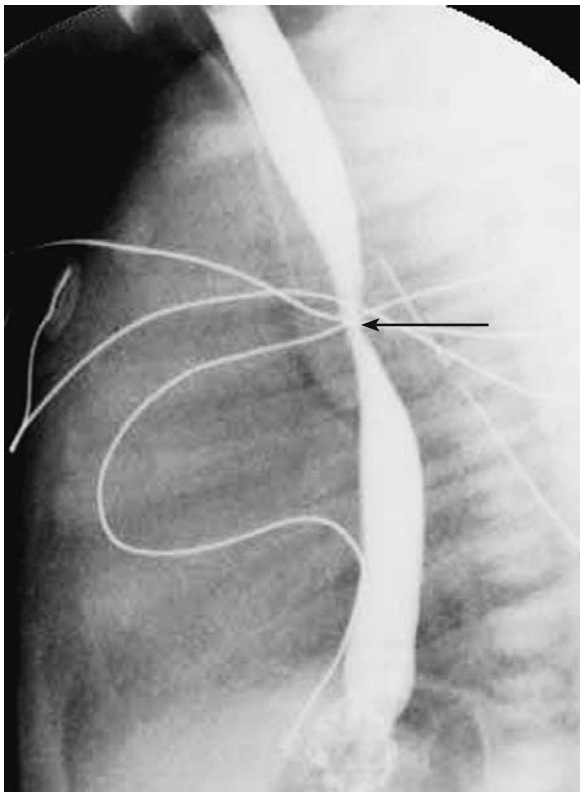


Fig. 2.12. Post-anastomotic narrowing of the esophagus. Routine post-operative swallow with non-ionic isotonic contrast medium shows esophageal narrowing at the primary anastomosis without evidence of hold up. No leak was demonstrated. Tracheomalacia is evident. Contrast medium in the airway is the result of an episode of aspiration (*arrow*)

2.4.3 Congenital Esophageal Stenosis

This is an uncommon anomaly, occurring in 1 in 25,000 to 1 in 50,000 live births (Murphy et al. 1995). Some consider esophageal stenosis to be a very mild variant of EA (Berrocal et al. 1999). The association with TEF in one-third of cases and other congenital anomalies in 17%–33% (Vasudevan et al. 2002) may support this theory. Most strictures are localized, 2- to 3-mm areas of narrowing approximately at the junction of the middle third and the distal one-third of the esophagus. Congenital esophageal stenosis may occur as an isolated narrowing due to ectopic cartilaginous tracheobronchial remnants, as an incomplete membranous diaphragm or web, or a localized segmental hypertrophy of the muscularis and submucosal layers with diffuse fibrosis (Murphy et al. 1995). There is a higher incidence of post-operative anastomotic leaks in children with TEF who have an associated congenital esophageal stenosis (Newman and Bender 1997). Esophageal stenosis usually presents when solid food is introduced or after impaction of a foreign body. In older children the differential diagnosis includes strictures due to reflux, caustic ingestion and sequelae from surgery. The diagnosis should be considered in an infant with dysphagia or with an impacted foreign body after EA repair. Esophageal stenosis must be considered in any child with acute dysphagia or foreign body impaction even without the antecedent history of atresia.

Treatment is resection with end-to-end esophageal anastomosis. Dilatation has a high incidence of esophageal perforation, especially in young children, and may be related to the length of the stricture and its transmural involvement (Newman and Bender 1997). The diameter of the lumen increases with age and growth of the patient, and with repeated dilatations, but will never be normal (Newman and Bender 1997). Some patients will benefit from initial dilatation, but most ultimately require surgery (Amae et al. 2003).

2.4.3.1 Radiological Findings

A congenital web appears as an oblique or transverse filling defect in the column of barium, and may be at the same level as a TEF. The esophageal narrowing is smooth and well defined, without evidence of ulceration. Barium swallow reveals an abrupt narrowing in the lower esophagus or a gradually tapering stenosis (Fig. 2.13) (Amae et al. 2003). Dilatation of the proximal esophagus reflects a high grade stenosis. Filling defects in the barium column can represent a foreign body or food bolus. The entire esophagus must be evaluated after surgical correction of EA. The contrast swallow may reveal an unsuspected congenital esophageal stricture (Vasudevan et al. 2002). The stenosis will be missed if the bolus of contrast medium is insufficient to cause adequate distention of the esophagus. Occasionally, a nasogastric tube is required to administer an adequate bolus to distend the entire esophagus. The esophagus must be evaluated dynamically during fluoroscopy to ensure that a stenosis is not missed or confused with spasm or peristalsis (Newman and Bender 1997).

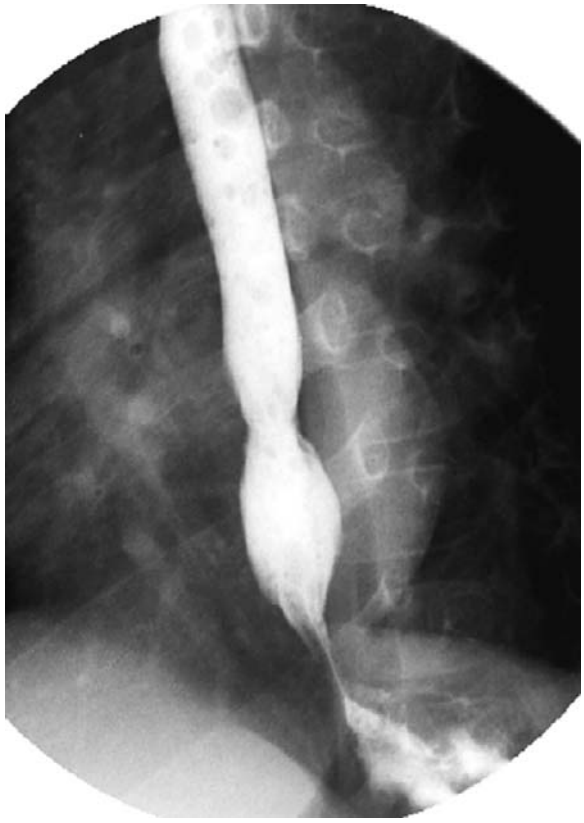


Fig. 2.13. Congenital esophageal stenosis. Barium swallow reveals a discrete annular narrowing in the distal esophagus. Tracheobronchial rests were found at pathology

2.4.4 Esophageal Duplication

The esophagus is the second most common location of duplication after the ileum, and accounts for 15%–20% of all duplications. It has been suggested that duplications result from aberrant luminal recanalization (Bremer 1944). In the 5^{th}–6^{th} week of intrauterine life, the foregut is covered by cells similar to those of the respiratory tract. The epithelium grows and obliterates the lumen, later producing secretions that form intercellular vacuoles that coalesce to form the new lumen. Failure of localized vacuole formation results in a cyst, which then migrates laterally into the esophageal wall and becomes surrounded by the muscular layer. Due to elongation of intrathoracic viscera, the cysts are commonly found in the lower esophagus. Complete esophageal duplication is extremely rare, and is often associated with gastric duplication (Singleton and King 1971; Herman et al. 1991). Esophageal duplication cysts are treated by excision.

2.4.4.1 Radiological Findings

Most esophageal cysts are detected incidentally on chest radiographs as mediastinal masses (Fig. 2.14). The esophagram demonstrates a well defined extrinsic soft tissue mass displacing the esophagus. The role of cross-sectional imaging is to differentiate the cyst from a solid mass such as neuroblastoma or sequestration and to show its relationship to adjacent vital structures. CT reveals a well defined fluid containing, non enhancing mass. Cyst contents have water-like characteristics on MR.

2.4.5 Esophageal Bronchus

Bronchopulmonary foregut malformations are rare congenital anomalies that are characterized by a fistula between an isolated portion of respiratory tissue and the foregut (Srikanth et al. 1992). Of these malformations, the esophageal bronchus communicating with the lower esophagus is the most common. Com-

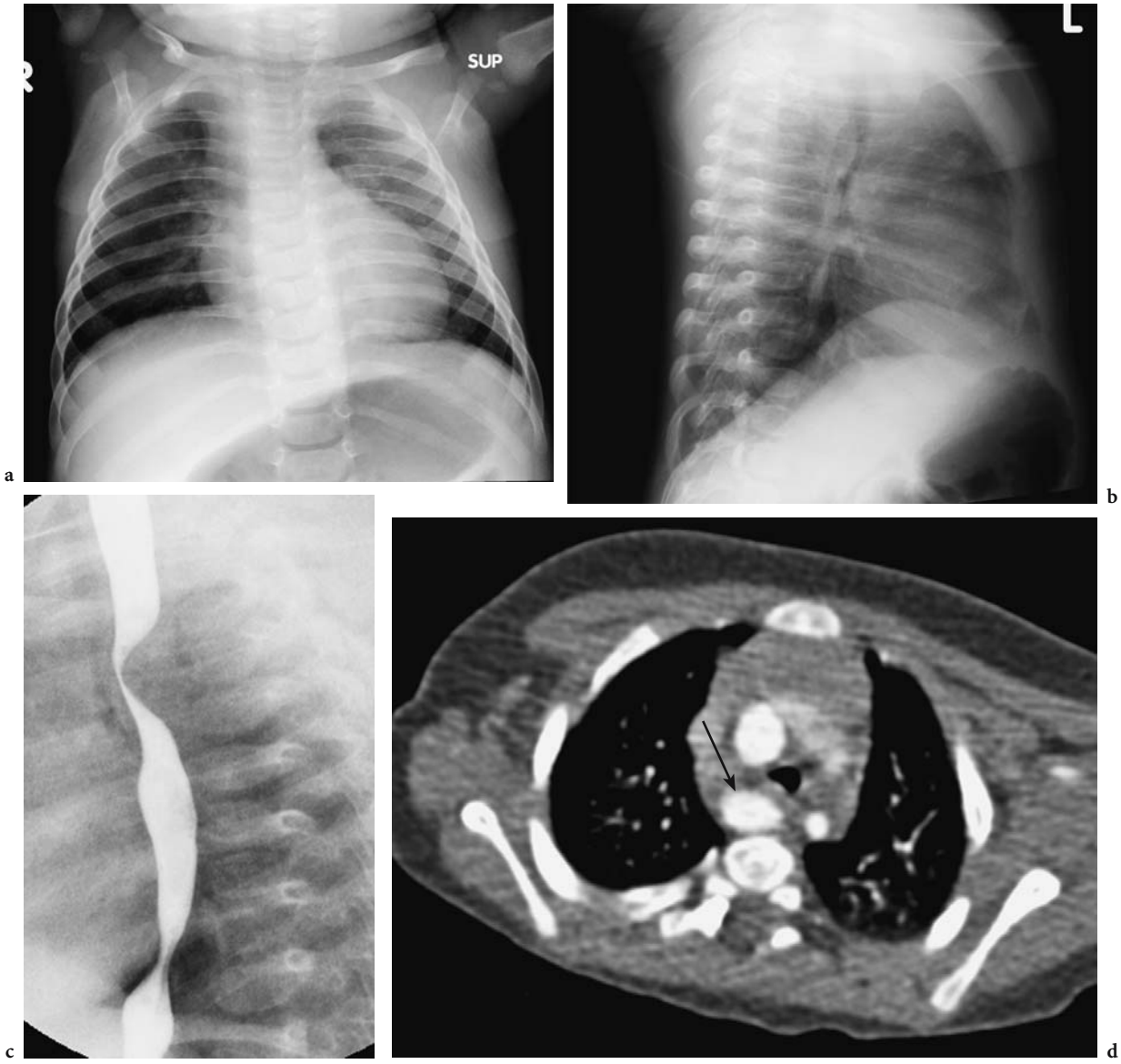


Fig. 2.14a–d. Esophageal duplication cyst. **a** AP chest radiograph without appreciable abnormality. **b** Lateral chest radiograph with anterior bowing and narrowing of the airway above the carina. **c** Barium esophagram confirms the presence of a soft tissue mass. **d** Contrast-enhanced CT shows the cyst posterior to the airway, and deforming the airway (*arrow*)

munication with the stomach is rarer. The abnormal communication develops when the lung sacs come into close contact with the esophagus and a part of the lung bud connects with the esophagus through a focal mesodermal defect. The attached portion of lung tissue, covered by mesenchyme, is carried away by the rapidly elongating esophagus (Srikanth et al. 1992). The arterial supply is variable and may arise from the pulmonary artery or the aorta and its branches. Venous drainage is commonly to the left atrium, but may also be systemic (Leithiser et al. 1985). This malformation is frequently associated with other anomalies. The abnormal connection to the respiratory tract causes respiratory distress, coughing with feeds and recurrent pneumonias.

Treatment is resection of the anomalous pulmonary tissue which is often hypoplastic and destroyed by infection. If detected early enough before pulmonary damage has occurred, tracheal reimplantation is preferred to resection (Michel et al. 1997).

2.4.5.1 Radiological Findings

The chest radiograph reveals opacification of the involved lung, and the communication is confirmed with a contrast examination of the esophagus and stomach (Fig. 2.15). Non-ionic isotonic contrast medium is the safest choice to minimize pulmonary complications such as acute pulmonary edema. Angiography is necessary to demonstrate adequate blood supply and venous drainage. CT will reveal signs of destroyed pulmonary parenchyma, which include atelectasis, bullae and abscesses. CT angiography (CTA) can demonstrate the source of arterial blood supply, its adequacy and venous drainage. Lung volume estimation can also be done with CT as part of the pre-surgery evaluation.

2.4.6 Vascular Ring Abnormalities

Vascular rings are uncommon. They are formed by vascular and ligamentous structures that encircle the airway and esophagus, causing extrinsic compression and obstruction of these hollow structures. Rings form as a result of developmental failure of parts of the paired 4^{th}–6^{th} aortic arches (Bonnard et al. 2003). Almost 20% of vascular ring malformations occur in association with congenital cardiac anomalies. Some rings are incomplete and asymptomatic, and are only incidentally discovered on chest radiographs or esophagrams. Symptoms vary with the degree of constriction of the vascular ring around the airway and esophagus. It is not surprising that the double aortic arch, which encircles the airway and the esophagus, causes the most severe symptoms. A double aortic arch is formed by a larger superior right aortic arch and a more inferior smaller left aortic arch. Symptoms from vascular rings are principally respiratory. Gastrointestinal symptoms and dysphagia are less common, and are more often encountered with a left retroesophageal subclavian artery arising from a right aortic arch (Bonnard et al. 2003). A right aortic arch with mirror-image branching acts like a vascular ring if the left ductus arteriosus passes between the right descending aorta and the left pulmonary artery.

The treatment is surgical relief of the constriction. The basic principle of vascular ring surgery is to divide non-functional or non-critical components of the ring. Surgical relief of the double aortic arch entails dividing the lesser of the two arches. An atretic portion is an optimal location for division of the arch. In cases with a right aortic arch and ligamentum arteriosum, the ductus remnant is divided, and the trachea and esophagus are released from adhesive bands (Backer et al. 2005).

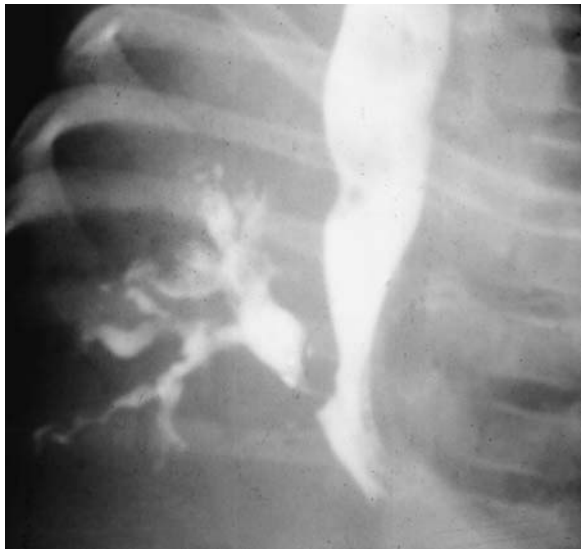


Fig. 2.15. Esophageal bronchus. Barium esophagram demonstrates that the right main bronchus arises from the esophagus. The right lung is completely opacified

2.4.6.1 Radiological Findings

The chest radiograph and esophagram demonstrate extrinsic esophageal impressions and tracheomalacia, as well as the side of the aortic arch. A right sided aortic arch in a child with respiratory symptoms is suggestive of a double aortic arch. The next examination is often a barium esophagram. A normal esophagram will clearly rule out the presence of a vascular ring. Symptomatic vascular rings caused by double aortic arches with bilateral arch patency cause significant narrowing and anterior bowing of the trachea, which are apparent on the lateral chest radiograph and barium esophagram. On the frontal projection, right and left indentations assume an "S" configuration. The barium esophagram is helpful to determine which component of a double aortic arch is larger. Barium swallow can show the right arch and a posterior esophageal impression (Fig. 2.16). This type of arch is usually associated with congenital heart disease. Advantages of the barium swallow include its ready availability, that

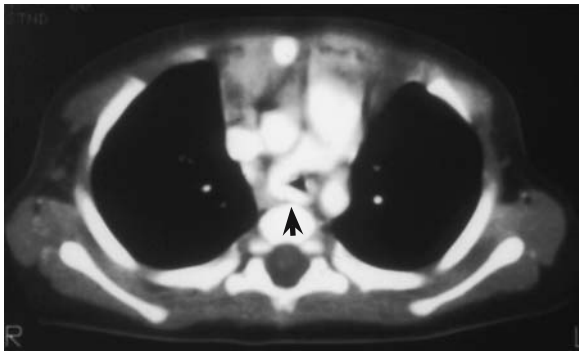


Fig. 2.16. Anomalous left pulmonary artery. CT with intravenous contrast medium shows the anomalous left pulmonary artery originating from the right main pulmonary artery, passing posterior to the trachea between the airway and the esophagus (*arrowhead*)

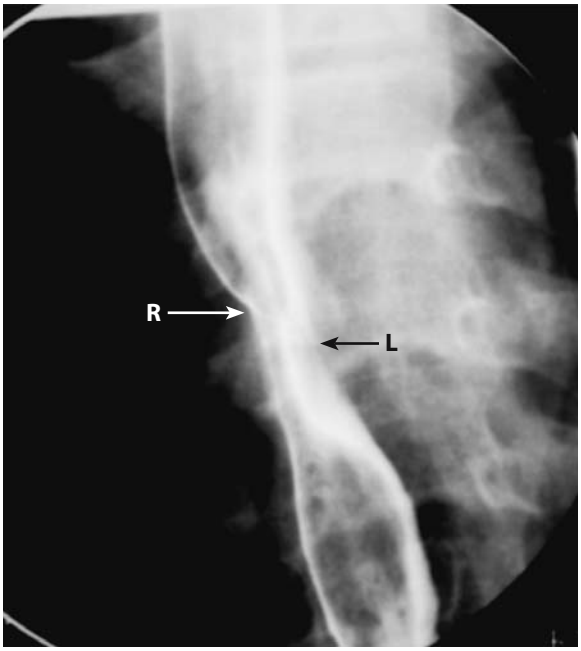


Fig. 2.17. Double aortic arch. Barium esophagram with double extrinsic impressions on the barium column. The dominant right arch (*R*) is higher than the smaller left arch (*L*)

it does not require sedation, an important feature for children who have airway compromise, and that it can show extrinsic compression by an atretic vascular segment not delineated by CT or MRI (Woods et al. 2001).

Over the last decade, however, the diagnosis of esophageal rings has changed from barium swallow and angiography to cross-sectional imaging (Figs. 2.17 and 2.18). CT angiography and MRI are optimal for more precise pre-operative delineation of the anatomy of the vascular ring (Hernanz-Schulman 2005). CT is more quickly performed, and can often be done without sedation. CT angiography can be completed in 20–30 s, whereas MR often requires sedation or immobilization for studies that can take as long as 45 min. The advantage of CT is visualization of the lungs and bronchi, and the diagnostic yield is even enhanced with multiplanar reformations. High-resolution 3D FISP (fast imaging with steady precession) MRA will accurately define vascular rings. This bright blood technique differentiates between vascular and non-vascular structures with higher spatial resolution than spin-echo (Greil et al. 2005). The choice of imaging modality varies with institutional preferences.

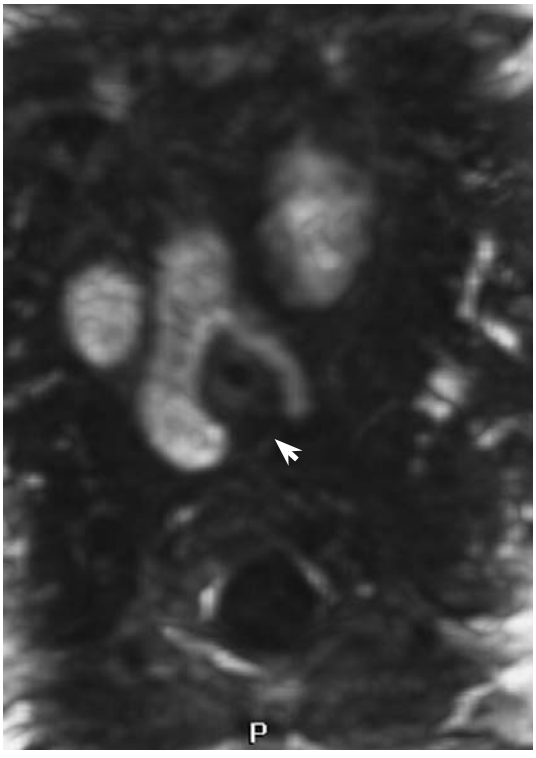


Fig. 2.18. Double aortic arch. MRI with contrast medium. The dominant right aortic arch and smaller left arch encircle the airway and the esophagus. The posterior limb of the vascular ring is atretic (*arrowhead*)

2.4.7 Hiatal Hernia

Hiatal hernia and intrathoracic stomach are uncommon in children. This condition may be congenital and inherited (Chana et al. 1996). Hiatal hernia may be secondary to congenital esophageal short-

ening (Chacko et al. 1998). Abnormally lax gastric ligamentous attachments and non-fixation of the stomach result in intrathoracic herniation in Marfan syndrome (Al-Assiri et al. 2005). Though usually an isolated anomaly, hiatal hernia can occur with other congenital abnormalities (Wang et al. 1993).

Esophageal clearance time and duration of gastroesophageal reflux are prolonged in children with hiatal hernia, thereby increasing esophageal exposure to acid and causing a higher failure rate of conservative therapy (Gorenstein et al. 2001).

2.4.7.1 Radiological Findings

Chest radiographs may reveal gas lucency in an intrathoracic stomach, occasionally with compressive atelectasis of adjacent lung. The next examination is the esophagram to confirm intrathoracic gastric malposition (Fig. 2.19). Gastric rugal folds of the hiatal hernia extend above the diaphragm or a portion of the gastric fundus protrudes through the esophageal hiatus. Sonography can demonstrate gastroesophageal reflux, as well as delineate the abnormally short length of abdominal esophagus and loss of the acute gastroesophageal angle that occur with a hiatus hernia (Westra et al. 1990).

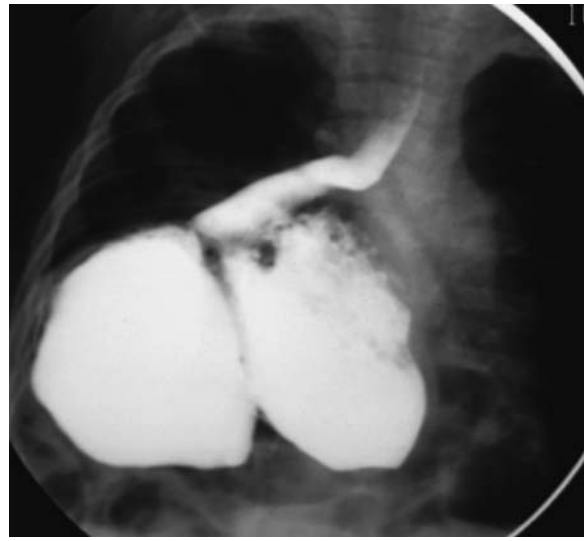


Fig. 2.19. Hiatal hernia. This infant with Marfan syndrome has intrathoracic gastric malposition

2.5 Acquired Abnormalities

2.5.1 Gastroesophageal Reflux

Gastroesophageal reflux (GER) is very common in infants and children, and may be physiological in young infants. The incidence ranges from 18% in all infants up to 70% in children with underlying conditions such as tracheoesophageal fistula, neurological deficits and anatomic abnormalities of the esophagus (McGuirt 2003). GER decreases spontaneously from an incidence of 67% at 4–5 months, declining to 21% by 6–7 months and to less than 5% by 12 months (Nelson et al. 1997). In young infants, the short length of the intraabdominal esophagus and physiologic immaturity of the developing lower esophageal sphincter contribute to GER, which invariably improves with the introduction of solid food. GER resolves spontaneously when most infants learn to sit up in the latter part of the first year of life, suggesting that gravity likely plays a role in aiding downward passage of esophageal contents. The presentation of GER is variable. In infants, GER manifests as regurgitation and "spitting up". Abdominal pain is a common symptom of GER in school-age children (Hassal 2005a).

There is a distinction between the common physiologic GER of childhood and pathological gastroesophageal reflux disease (GERD). This more severe form of reflux can interfere with growth, and cause gastroesophageal and respiratory symptoms. GER has been linked to asthma, and pulmonary symptoms are significantly higher in children with GER than those without (Gold 2005). Children and adolescents with GER are more likely to present with cough and other respiratory symptoms than complaints of "heartburn". Asthma itself causes GER by a variety of mechanisms. Hyperinflation changes the pressure gradient across the lower esophageal sphincter, increases negative intrathoracic pressure and alters the relationship between the diaphragm and lower esophageal sphincter. This may be exacerbated by some asthma medications that decrease lower esophageal sphincter pressure.

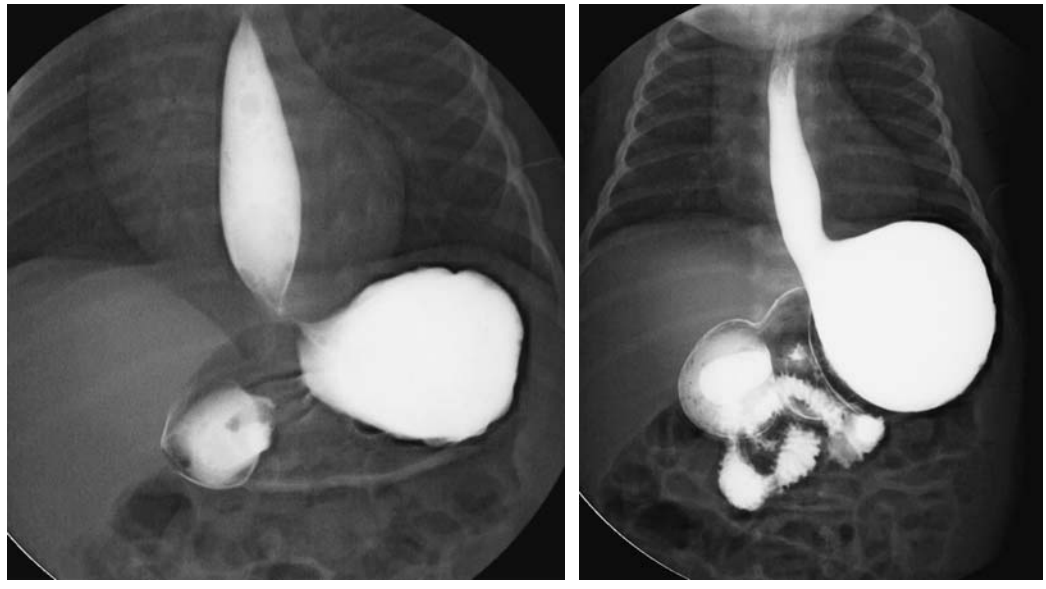
While most children with "physiologic" GER will naturally outgrow the reflux, those children with underlying abnormalities will not (Boix-Ochoa and Canals 1976). The initial treatment for GER is most commonly thickening of the infant's formula, feeding smaller amounts per meal and maintaining the

infant in an upright position after each feed. Esophageal pH monitoring is the standard and reliable method to document abnormal gastric acid reflux, as well as to assess the efficacy of therapy in patients who do not respond to acid suppression treatment (Rudolph et al. 2001). Early diagnosis and treatment will prevent and mitigate complications such as failure to thrive, refusal to feed and respiratory disorders. Children above 3 years with GER have a higher rate of related complications and frequently require medical or surgical intervention (McGuirt 2003). Fundoplication augments the lower esophageal sphincter with a wrap of the gastric fundus; it is the most common surgical treatment for GER, but has a high rate of failure, which ranges from 30%–70% within 1–3 years (Hassal 2005b). Ironically, those with the greatest need for good reflux control with neurologic impairment, repaired esophageal atresia, chronic lung disease have underlying pathophysiological mechanisms that lead to wrap failure (Hassal 2005a).

2.5.1.1 Radiological Findings

GER is the commonest indication for performing barium esophagram and upper gastrointestinal series in children. To optimize the examination, the stomach should be filled with the same volume as with a normal feed. If the child refuses to drink sufficient barium, the ingested volume can be increased with formula or fruit juice. The volume can be supplemented with fluid injected through an indwelling gastrostomy tube, or the stomach can be filled through a nasogastric tube which is removed after filling. Nasogastric tubes maintain patency of the lower esophageal sphincter and compromise its function. Tubes must be removed when evaluating for reflux. The barium swallow has only 50% sensitivity and specificity for reflux diagnosis in children (Rudolph et al. 2001). Reflux of barium is not diagnostic of GERD, and nor does absence of reflux rule it out (Rudolph et al. 2001). The most important aspect of the barium examination is to exclude anatomic abnormalities of the esophagus, to define the level of the duodenojejunal junction and to define the cephalad anatomic level of reflux, as well as to document aspiration (Fig. 2.20). Aspiration with reflux may be seen, but is very uncommon (Fernbach 1994). A carefully performed upper GI series may miss significant GER because of the limited use of fluoroscopic monitoring time and the relatively short duration of the entire examination.

The radionuclide "milk scan" is a sensitive test for diagnosing GER (Blumhagen et al. 1980; Seibert et al. 1983). Milk, formula or juice mixed with Tc 99m sulphur colloid is administered to the child who is then scanned. Radionuclide scanning is continuous, an advantage over fluoroscopy. Radionuclide scanning allows documentation of episodes of GER, and



a b

Fig. 2.20a,b. Gastroesophageal reflux. **a** During swallowing, the gastroesophageal junction is closed. **b** The gastroesophageal junction is widely patent, and barium refluxes to the upper esophagus

cephalad extent of GER, as well as the rate of gastric emptying.

GER can be demonstrated sonographically into the distal esophagus (Koumanidou et al. 2004). Gastric contents can be observed as they reflux into the esophagus. This technique is limited because the degree of reflux and proximal extent cannot be evaluated in the chest where the esophagus is obscured by the lungs.

2.5.2 Achalasia

Achalasia is rare in children with an incidence estimated at approximately 0.05–1 per 100,000. Fewer than 5% of those with achalasia present in childhood (Emblem et al. 1993; Mayberry and Mayell 1998). This condition is characterized by defective relaxation of the cardia and absence of esophageal peristalsis with normal upper esophageal sphincter and pharyngeal function. Pathological findings are characterized by marked fibrotic hypertrophy of the myenteric plane between the muscle layers and a significant reduction in the number of myenteric ganglia and myenteric neurons (Khelif et al. 2003). It is not uncommon for children with achalasia to present before the age of 5 (Hussain et al. 2002). Clinical symptoms are age-related. Infants present with symptoms similar to gastroesophageal reflux, including frequent regurgitation, choking, apnea and pneumonia. Symptoms in older children are similar to those of adults and include dysphagia, chest pain, cough, vomiting of undigested food and poor weight gain (Vane et al. 1988). As a result of its rarity in children and non-specific symptomatology, the diagnosis and treatment are frequently delayed.

Injection of botulinum toxin temporarily relieves the symptoms of achalasia, and half of the patients treated with botulinum toxin will require an additional procedure. Botulinum toxin is only recommended for patients who are poor candidates for pneumatic dilatation or surgery (Hurwitz et al. 2000). Pneumatic dilatation has a higher success rate and lower failure rate than botulinum toxin. Modified Heller myotomy has 94% efficacy and pneumatic dilatation has 90% efficacy (Vaezi and Richter 1999). Myotomy alone has a high incidence of gastroesophageal reflux (Emblem et al. 1993), and some children ultimately require an antireflux procedure (Vane et al. 1988). Laparoscopic modified Heller myotomy is increasingly more commonly performed in children and can be performed in patients as young as 10 years. The myotomy relieves symptoms of obstruction, yet the underlying esophageal dysmotility persists through the patient's life. These patients must be monitored and followed up for life.

2.5.2.1 Radiological Findings

The chest radiograph may reveal a dilated esophagus with an air fluid level, changes of chronic aspiration and tracheal displacement (Fig. 2.21). The stomach

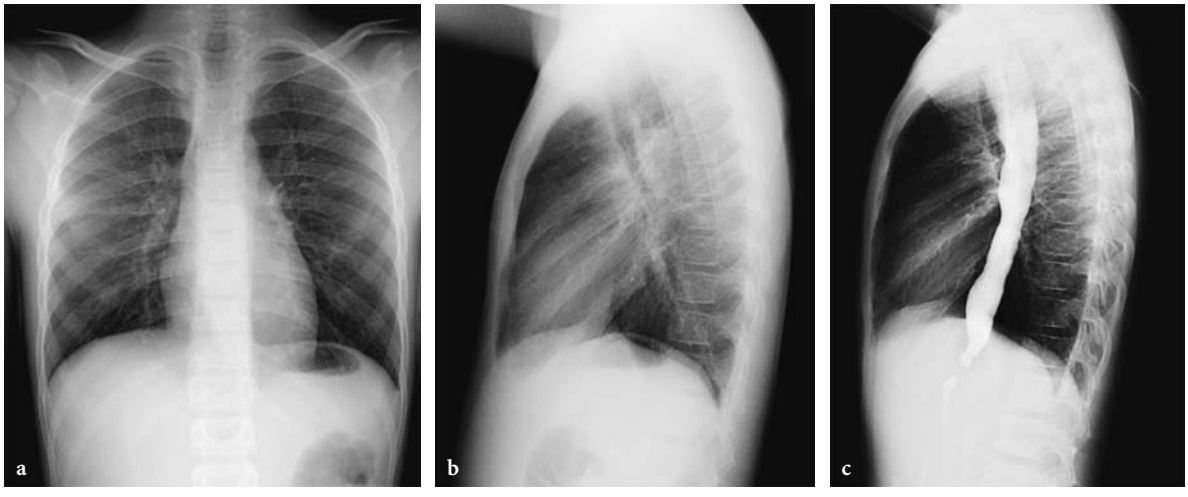


Fig. 2.21a–c. Achalasia. AP (**a**) and lateral (**b**) chest radiographs reveal an air-fluid level in the distended esophagus. **c** Delayed lateral chest radiograph. Barium fills the entire length of the esophagus, which tapers distally. There has been minimal flow of barium into the stomach

bubble is not visualized in cases with high grade obstruction. The esophagram is the initial diagnostic study, followed by esophageal manometry and endoscopy. Barium esophagram reveals a dilated esophagus that tapers smoothly distally to a "bird's beak". Occasionally, a leiomyoma may mimic the symptoms and radiographic appearance of achalasia (Hussain et al. 2002).

2.5.3 Foreign Body Ingestion

Infants and young children experiment with unfamiliar objects by placing them in their mouths. Most swallowed foreign objects pass uneventfully. The age range of children who swallow foreign bodies is from 6 months–3 years. A higher incidence of obstructed ingested foreign bodies occurs with esophageal pathology, especially after repaired esophageal atresia and among children with psychiatric disease and retardation. Foreign bodies are most likely to be caught at the normal anatomic sites of esophageal narrowing. Foreign bodies at other levels are indicative of a stricture or vascular ring (Fig. 2.22). Foreign bodies lodge at the level of the thoracic inlet (53%) in the thoracic esophagus (32%), and the least common site is the cervical esophagus (15%) (Harned et al. 1997). Presentation is usually with dysphagia and chest pain. Salivation and drooling occur with esophageal obstruction. Swallowed objects may be unchewed or partially chewed food, or other foreign objects, 66% of which are coins in children (Fig. 2.23) (Webb 1995).

More than one coin may occasionally be ingested (Fig. 2.24). The older the children, the larger the coins ingested (Cheng and Tam 1999). Acute coin ingestion is rarely symptomatic unless the coin is above the thoracic inlet (Sharieff et al. 2003). The majority of coins will not pass spontaneously. However, some coins will pass spontaneously and can be followed for 24 h, thereby decreasing the need for operative removal (Sharieff et al. 2003). The coin should be removed if repeat radiography reveals non-passage. Flexible endoscopy is safer than blind removal, and provides immediate information about the esophagus at the site of impaction.

Button battery ingestion is rare, but the incidence is increasing (Yardeni et al. 2004). Most are less than 15 mm in diameter, and pass uneventfully. Button batteries contain alkali and cause injury by direct corrosion, voltage burns and pressure necrosis. Those that lodge can cause severe complications within a short time after ingestion and must be removed endoscopically as soon as possible.

Complications of foreign body ingestion include perforation and abscess formation. Flexible endoscopy is the most common method for removal.

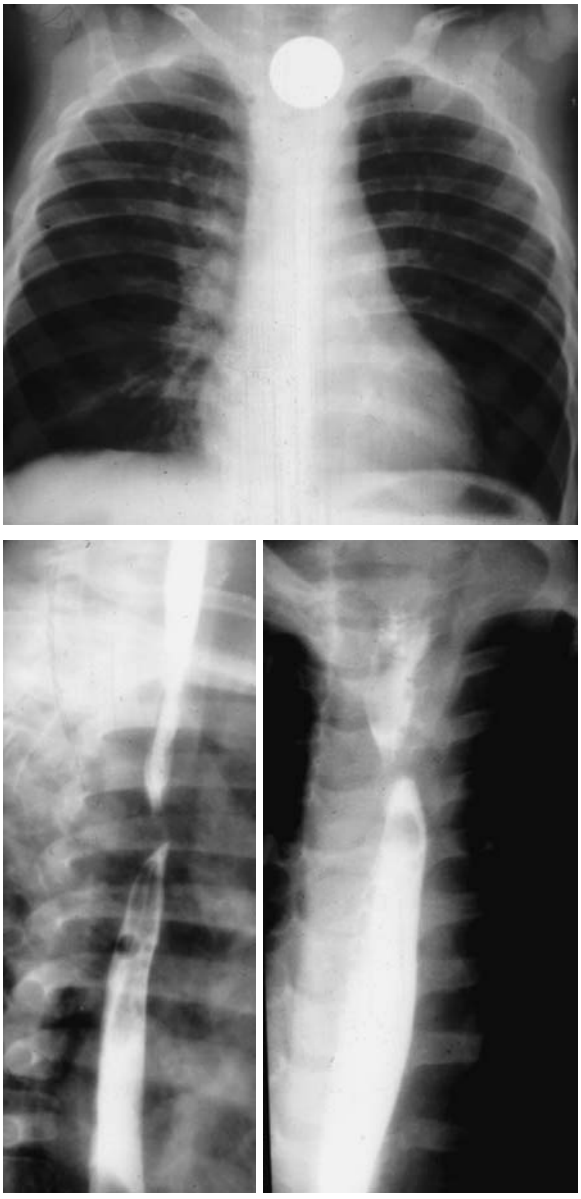


Fig. 2.22a,b. Impacted coin above an unsuspected vascular ring. **a** Frontal chest radiograph shows the coin lodged in the proximal esophagus above the aortic arch. **b** Barium esophagram reveals an aberrant retroesophageal right subclavian artery

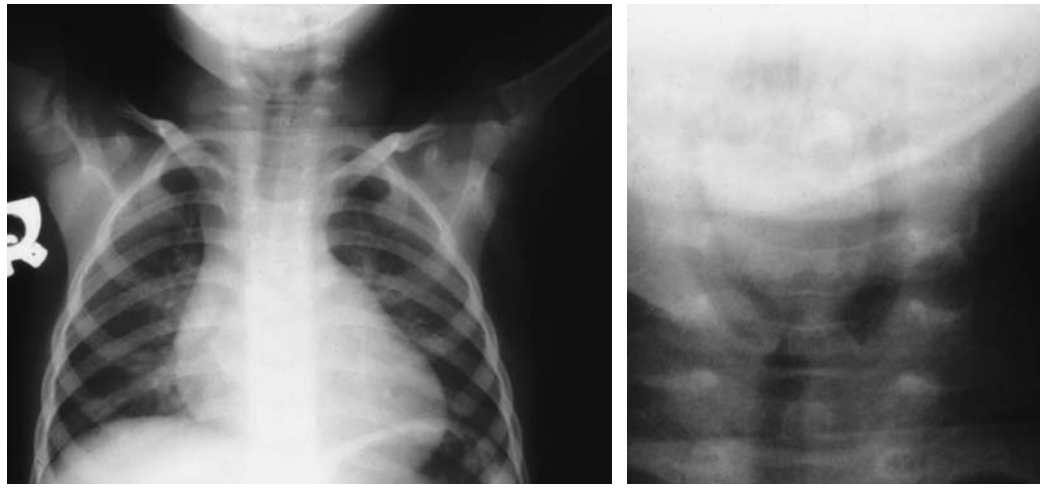


Fig. 2.23a,b. Radiolucent foreign body. AP (**a**) chest radiograph and magnified view (**b**) reveal an impacted piece of unchewed chicken in the cervical esophagus outlined by esophageal air

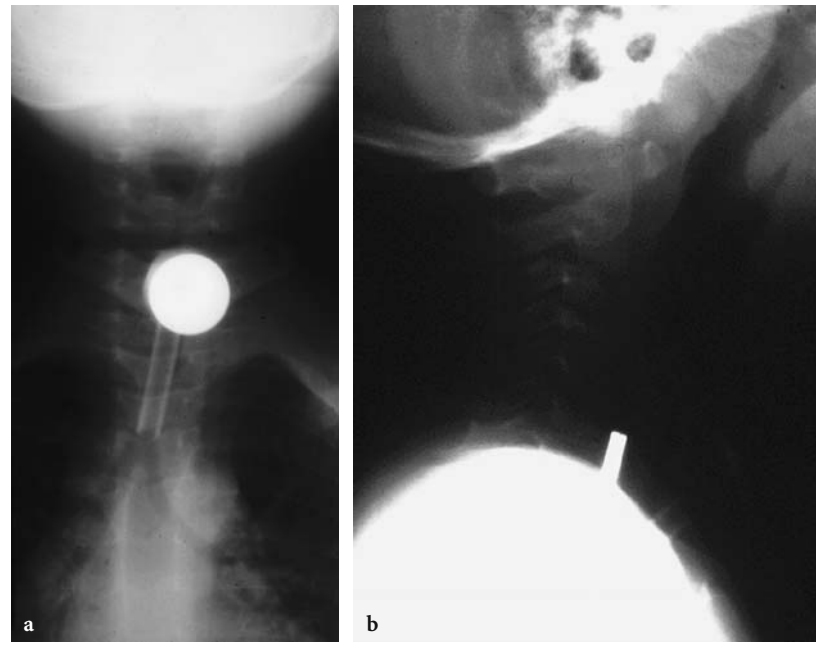


Fig. 2.24a,b. Two impacted coins. **a** The AP view of the airway is confusing. The metallic density resembles part of the tracheostomy tube. **b** The lateral radiograph reveals two coins in the esophagus

2.5.3.1 Radiological Findings

When suspicion for an ingested foreign body is high, frontal and lateral radiographs should be obtained from the nasopharynx to the abdomen. Radiology reveals 100% of metal objects, 86% of glass and 26% of fish bones (Cheng and Tam 1999). Commonly ingested foreign bodies such as medications, small plastic toys and organic material are not seen on plain radiography. These radiolucent foreign bodies may show on the barium esophagram as filling defects in the barium. A contrast examination must not be performed with a high grade obstruction because of the aspiration risk. Chronically lodged foreign bodies cause inflammation, edema of the esophageal wall and narrowing of the lumen (Fig. 2.25).

Fluoroscopic Foley catheter removal has a high (91%) success rate with coins that have been in place for less than 3 days. The rate of successful removal is lower with coins that have been lodged for a longer time period (Schunk et al. 1994). The success rate is

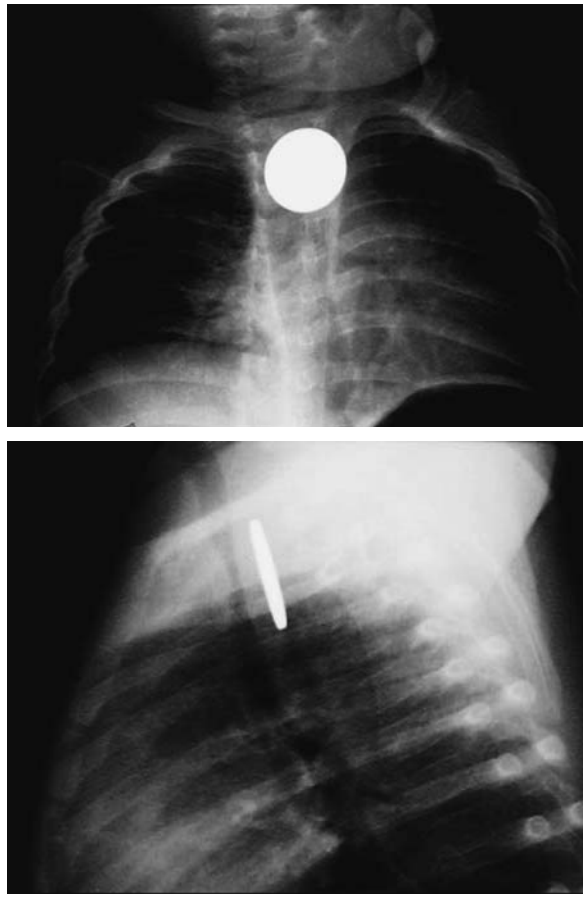


Fig. 2.25a,b. Chronically impacted coin. Frontal (**a**) and lateral (**b**) chest radiographs with an impacted coin and inflammatory narrowing of the adjacent airway. The space between the trachea and the esophagus is thickened, evidence of chronic inflammatory change

also lower (83%) in cases with underlying esophageal pathology (Schunk et al. 1994). An underlying stricture will not permit passage of the Foley catheter, and may be a cause for failure. Foley catheter removal is limited to objects without sharp edges. This technique must not be attempted in children with clinical or radiological airway compromise and should not be attempted in the presence of esophageal edema (Schunk et al. 1994).

Patients for fluoroscopic removal should not be sedated in order to maintain their airway. However, patients must be restrained and placed in the prone oblique position. A Foley catheter size 8–12 is placed through the nose or mouth under fluoroscopy to below the foreign body. The balloon is then inflated with 3–5 ml of contrast medium, taking care not to over distend the esophagus. The catheter is gently withdrawn, and the foreign body is delivered into the hypopharynx from where it can either be spontaneously expectorated, or manually removed by the radiologist whose fingers are in the child's mouth. A useful tip is to don two pairs of rubber gloves as these offer protection from being bitten by the patient. Occasionally, the Foley catheter will push the foreign body distally into the stomach. Complications are minor, and may include epistaxis and vomiting, but esophageal laceration may occur (Schunk et al. 1994). Although effective, this procedure is not widely performed and many pediatric radiologists defer to endoscopic extraction.

2.5.4 Caustic Ingestion

Caustic ingestion is rare in children. Most are accidental, and 58% occur in children younger than 6 years (Duncan and Wong 2003). Ingestion may be acid or alkaline products. Injury depends on ingestant characteristics, i.e. corrosive properties, the amount and concentration as well as the physical form of the substance, as well as the duration of mucosal contact. Alkalis initially cause liquefactive necrosis, and the most severe type of caustic injury, followed by scar formation and strictures. Most damage occurs in the middle and lower esophagus. Alkali granules cause strictures at any level where they may lodge. The esophagus tends to be spared injury in acid ingestion because acids, which tend to be liquids, pass through the esophagus more quickly than alkalis. Acids require longer contact to cause deep tissue injury and it is the stomach that sustains most injury.

Treatment is initially with volume resuscitation and airway management. Sips of water are encouraged to dislodge lye particles. Emetics and neutralizing agents are contra-indicated because heat damage can occur during neutralization, and emesis of acid gastric contents can damage the esophageal mucosa. Surgery is rarely required. Colonic interposition or gastric pull up are performed if the stricture is long and does not respond to dilatation (Fig. 2.26).

2.5.4.1 Radiological Findings

Chest and abdomen radiographs are obtained to screen for signs of perforation. The initial radio-

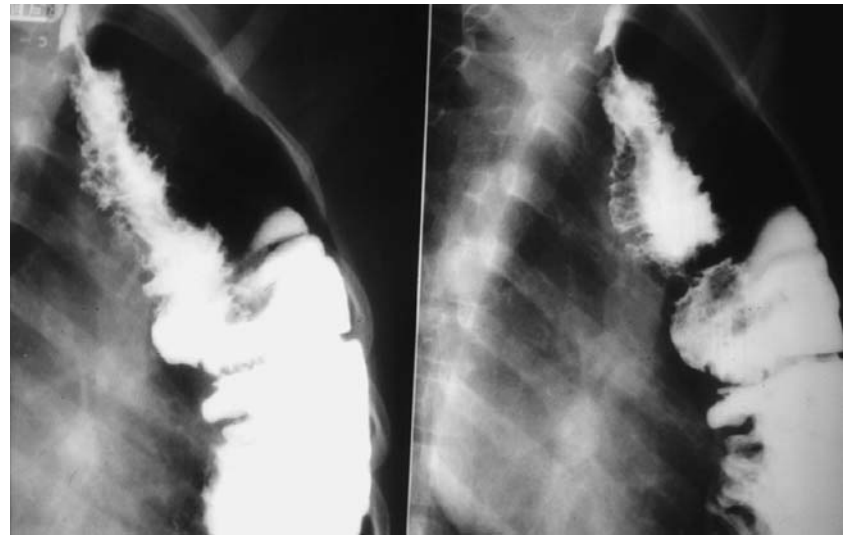


Fig. 2.26. Alkali ingestion. Barium swallow shows a long segment of narrowing of the mid esophagus approximately 3 weeks after swallowing drain cleaner

logic examination is the esophagram which is best performed with water soluble non-ionic isotonic contrast medium in case of perforation. The earliest signs are epiglottic swelling, esophageal dysmotility, mucosal edema and ulceration (Fig. 2.27). Intramural contrast and persistent gaseous dilatation of the esophagus are signs of severe injury and may precede perforation (Fernbach 1994). CT is recommended if suspicion for perforation is high. Deep esophageal burns are investigated by serial esophagrams to detect early stricture formation. Follow up barium esophagrams are important to evaluate the development of scarring and the length of the resulting strictures which are typically long.

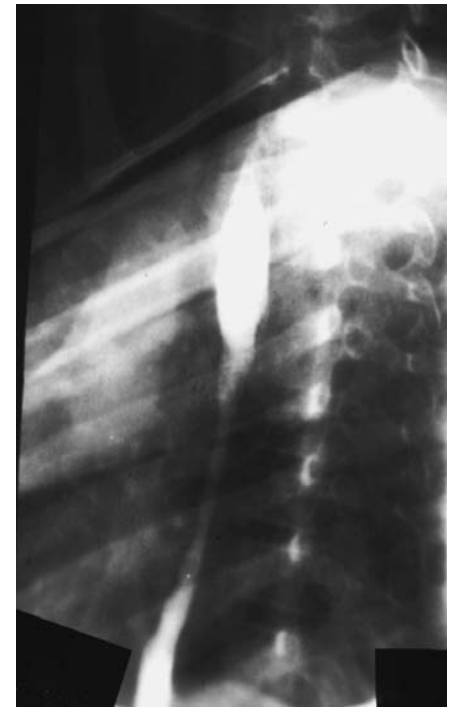


Fig. 2.27. Colonic interposition. Treatment for a high-grade long segment caustic stricture

2.5.5 Esophageal Strictures

Acquired strictures in children are most commonly encountered after surgical repair of EA and TEF. Strictures in the pediatric population may be caused by caustic ingestion, esophagitis and epidermolysis bullosa. Symptoms vary with the degree of tightness of the stricture, and include dysphagia, chest pain, cough and vomiting of undigested food. Drooling and refusal to eat may be signs of a foreign body causing complete obstruction of the stricture.

Balloon dilatation is the preferred treatment above traditional bougienage, and surgery is rarely indicated.

2.5.5.1 Radiological Findings

The chest radiograph is usually normal. Occasionally, an esophageal air-fluid level can be seen above an esophageal obstruction. Barium esophagram, the modality of choice, demonstrates strictures as nar-

rowing of the esophageal lumen and lack of distensibility which may be localized or diffuse (Fig. 2.28) (Karasick and Lev-Toaff 1995). The radiological appearance varies with the type of stricture and its caliber.

Fluoroscopic dilatation has many advantages over bougienage because balloon dilatation is not limited by the diameter of the nose or pharynx, and the incidence of perforation with balloon dilatation is much lower than with bougie dilatation (Fasulakis and Andronikou 2003). Serial balloon dilatation is recommended because progressive stretching of scar tissue prevents tears and perforations. Scar tissue can limit success since fibrosis and altered blood supply reduce tissue elasticity (Fasulakis and Andronikou 2003). The balloon is inflated under fluoroscopy at the level of the stricture, applying uniform radial force that is less traumatic than the shearing force of bougienage. Fluoroscopy has the added advantage of allowing the radiologist to check that the stricture is dilated to a suitable diameter. After dilatation, the success of the procedure can be monitored immediately with the introduction of water soluble contrast medium to show an increase in esophageal caliber and to evaluate for a leak or perforation (Fasulakis and Andronikou 2003).

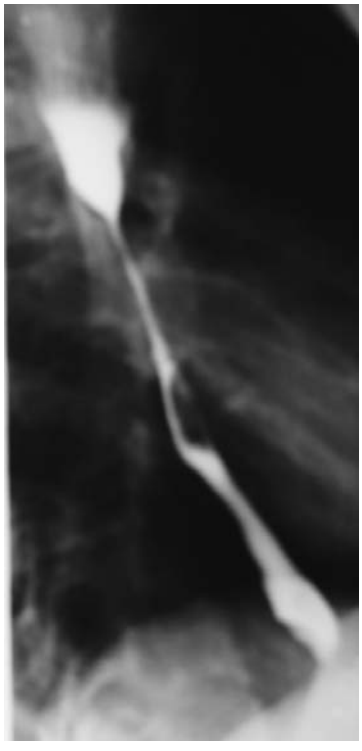


Fig. 2.28. Esophageal stricture. Barium swallow shows a long stricture involving almost the entire length of the esophagus years after ingesting an unknown corrosive agent

Although strictures may not be completely resolved after balloon dilatation, the procedure will provide functional relief, often further aided by forceful swallowing by the patient (Allmendinger et al. 1996). Balloon dilatation has to be repeated as the child grows (Allmendinger et al. 1996). Ongoing and progressive diseases such as repaired esophageal atresia and epidermolysis bullosa require repeated dilatation as the disease progresses (Fig. 2.29).

2.5.6 Esophageal Perforation

Esophageal perforations are rare in children, but the incidence is increasing as more diagnostic and therapeutic endoscopies are performed. Iatrogenic esophageal perforation is the cause in 33%–75% of cases (Martinez et al. 2003). The incidence is low in upper endoscopy, and higher with rigid dilators. Esophageal perforations are more likely to occur if a foreign body has been present more than 24 h and caused pressure necrosis. Other etiologies are pill-induced, caustic damage, infectious, including candida, herpes and tuberculosis. Cervical esophageal perforation may result from penetrating trauma by objects in the mouth, including lollipops and pencils. Esophageal perforation is potentially life-threatening because it allows entry of bacteria and digestive enzymes into the pleural and subphrenic spaces and the mediastinum, causing sepsis. Perforation of the intraabdominal esophagus may lead to sepsis and shock.

Conservative non-operative therapy for esophageal perforation is preferred. In children this consists of antibiotic coverage, drainage of pleural effusions, esophageal rest and total parenteral nutrition if there is no evidence of contrast leak at esophagram. Successful outcome depends on early diagnosis and treatment, young age and absence of underlying disease (Martinez et al. 2003). Operative treatment may be required with esophageal perforation or gross leakage.

2.5.6.1 Radiological Findings

Esophageal perforation can be diagnosed on frontal and lateral chest radiographs. Findings include pneumomediastinum, pneumothorax, hydro-pneumothorax, subcutaneous emphysema and pleural effusions (Fig. 2.30). Chest radiography is not useful

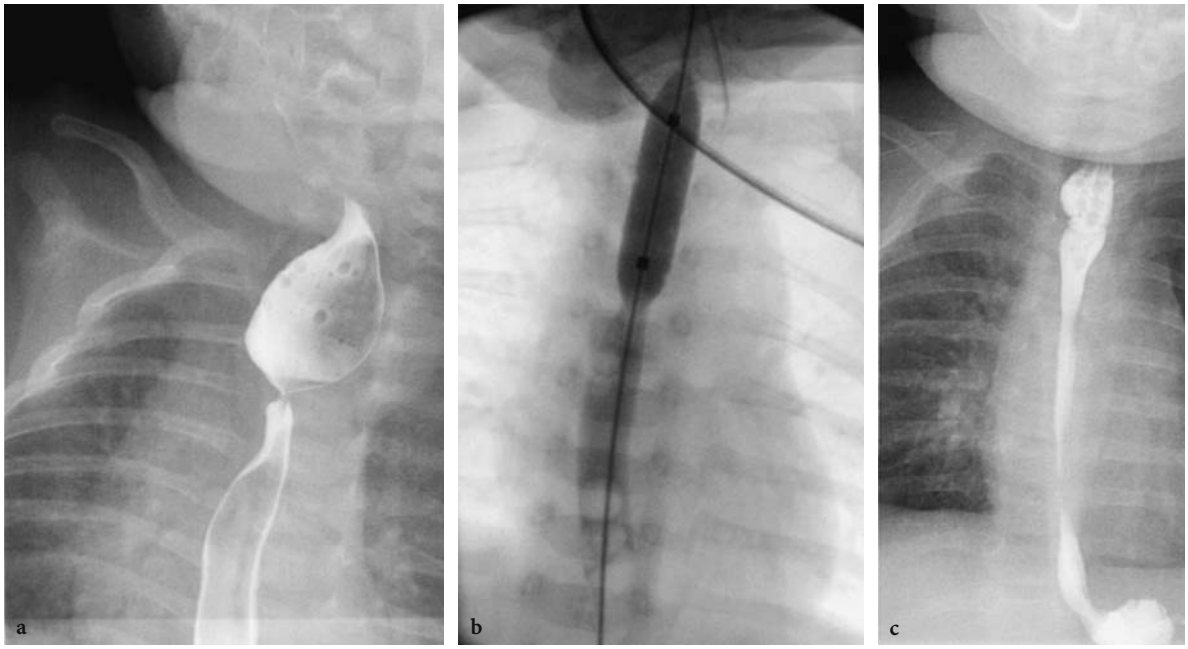


Fig. 2.29a–c. Balloon dilation of an esophageal stricture. **a** Severe stenosis of the proximal esophagus 3 months after repair. **b** Dilatation balloon expanded across the narrowing, no extravasation. **c** Post-dilatation appearance

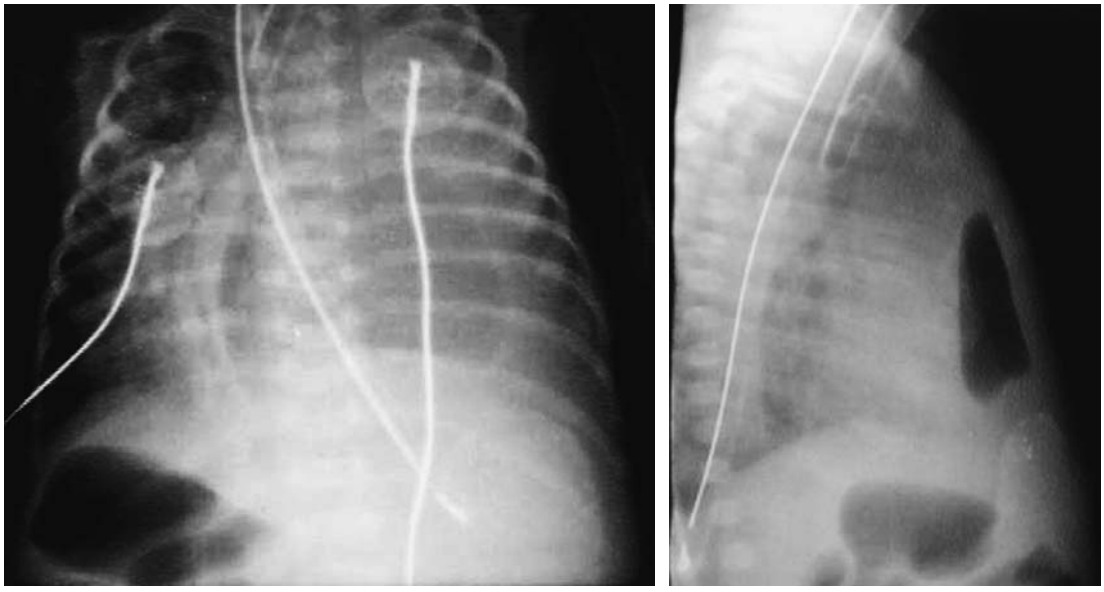


Fig. 2.30a,b. Esophageal perforation after orogastric tube malposition. **a** Frontal chest radiograph with left pleural effusion, left pulmonary atelectasis and mediastinal shift. Free air overlies the cardiac silhouette. **b** Lateral chest radiograph confirms the traumatic pneumothorax

with cervical esophageal perforation. Airway films are required if there is concern for cervical esophageal perforation. Contrast swallow is falsely negative in 10% (Gimenez et al. 2002). CT is recommended when the esophagram is negative and there is high suspicion of perforation.

2.5.7 Epidermolysis Bullosa

Epidermolysis bullosa is a rare inherited genodermatosis resulting from a mutation in COL7A1, the gene encoding type VII collagen (Horn and Tidman 2002). Clinical manifestations are caused by the extreme vulnerability of squamous epithelium of the skin and mucous membranes to minor trauma. The most affected subtype is the recessive epidermolysis bullosa dystrophica, in which the most severely affected individuals will develop blisters within the first 24 h of life (Horn and Tidman 2002). Most cases with esophageal involvement become symptomatic during the first two decades of life. Gastrointestinal manifestations can occur in the absence of active blistering (Horn and Tidman 2002). Swallowing is sufficiently traumatic to cause bullae and erosions that rupture, ulcerate and scar. Oral changes compromise the child's ability to chew food properly and further exacerbate the esophageal trauma (Anderson et al. 2004). Scarring and fibrosis cause shortening of the esophagus that may result in gastroesophageal reflux and further stricturing (Anderson et al. 2004).

Epidermolysis bullosa is an ongoing and progressive disease and repeated dilatations are necessary (Demirogullari et al. 2001). Dilatation is best performed in the inactive stage of the disease rather than during the active stages of blistering.

2.5.7.1 Radiological Findings

Affected individuals have a 15% incidence of developing squamous carcinoma at an early age (Hoeffel et al. 1992). Therefore, fluoroscopy must be prudently limited in these children and repeated fluoroscopy and fluoroscopically guided esophageal dilatation are discouraged

Children with epidermolysis bullosa must be treated very carefully to prevent trauma to their extremely fragile skin and mucosa. They should lie on soft padding on the fluoroscopic table and should not be restrained for radiographic procedures. Affected children should drink the barium spontaneously, and nasogastric tubes must be avoided (Fordham 2005). Barium esophagram will demonstrate strictures, approximately half which are in the proximal third of the esophagus near the cricopharyngeus muscle, 25% in the distal one-third, and the remainder in multiple sites (Fig. 2.31) (Kern et al. 1989). The strictures may be several centimeters in length, and are likely exacerbated by gastroesophageal reflux. Annular strictures are usually less than 3 cm in length.

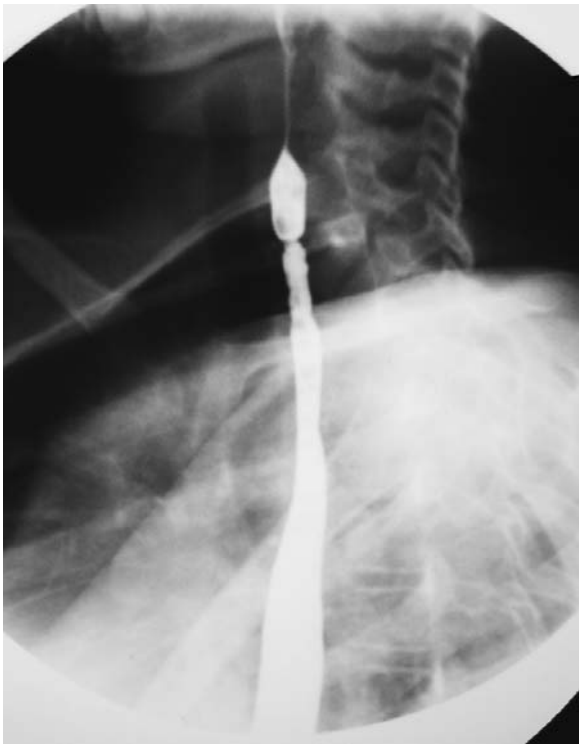


Fig. 2.31. Epidermolysis bullosa. Barium swallow reveals a proximal annular stricture and a long segment of diffuse irregularity of the esophagus

2.5.8 Infectious and Inflammatory Conditions

2.5.8.1 Infectious Esophagitis

Infectious esophagitis is rare in children. Candida is the most common cause, and usually occurs

in immunocompromised patients (Levine and Rubesin 2005). Candida esophagitis is common in the absence of oral thrush. Barium esophagram demonstrates linear or irregular filling defects separated by normal mucosa (Levine and Rubesin 2005). Herpes and cytomegalovirus are other frequent causes of esophagitis in immunosuppressed patients. They manifest on barium swallow as discrete well defined areas of ulceration (Levine and Rubesin 2005). HIV infection itself can cause large esophageal ulcers (Sor et al. 1995).

2.5.8.2 Inflammatory Esophagitis

Eosinophilic esophagitis is an atopic condition in which esophageal inflammation occurs with a predominantly eosinophilic infiltrate that extends into the muscularis. The annual incidence is 1 in 10,000, and is increasing (Noel and Rothenberg 2005). Symptoms mimic reflux esophagitis, are unresponsive to acid suppression therapy and respond to steroids. The esophagram shows a narrow esophageal lumen, caused by thickening of the esophageal wall, a feature also demonstrated on CT (Sant'Anna et al. 2004).

2.5.9 Esophageal Varices

Esophageal varices in children are a manifestation of portal hypertension that causes hepatofugal flow through esophageal collateral veins to drain into the superior vena cava. The most common children's diseases that cause portal hypertension include umbilical venous catheterization, biliary atresia, alpha-1 antitrypsin deficiency, autosomal recessive polycystic renal disease and cystic fibrosis.

2.5.9.1 Radiological Findings

On barium swallow, varices are demonstrated as serpiginous filling defects, best seen in the collapsed esophagus. The normal parallel course of the mucosal folds is interrupted by the varices that appear as filling defects. At the present time, barium swallow is uncommonly performed to make this diagnosis. Instead, the diagnostic work up includes Doppler sonography, MR cholangiography and MR angiography.

References

Al-Assiri A, Wiseman N, Bunge M (2005) Prenatal diagnosis of intrathoracic stomach (gastric herniation). J Pediatr Surg 40:E15–E17

Allmendinger N, Hallsey MJ, Markowitz SK et al (1996) Balloon dilatation of esophageal strictures in children. J Pediatr Surg 31:334–336

Amae S, Nio M, Kamiyama T (2003) Clinical characteristics and management of congenital esophageal stenosis: a report on 14 cases. J Pediatr Surg 38:565–570

Anderson SHC, Meenan J, Williams KN et al (2004) Efficacy and safety of endoscopic dilatation of esophageal strictures in epidermolysis bullosa. Gastrointest Endoscop 59:28–32

Babu R, Pierro A, Spitz L et al (2000) The management of oesophageal atresia in neonates with right-sided aortic arch. J Pediatr Surg 35:56–58

Backer CL, Mavroudis C, Rigsby CK, Holinger LD (2005) Trends in vascular ring surgery. J Thorac Cardiovasc Surg 129:1339–1347

Bannister LH (1995) Chapter 12. The alimenatary system. In: Banister LH, Berry MM, Colllins P et al. (eds) Gray's anatomy, 38th edn. Churchill Livingstone, Edinburgh, pp 1683–1812

Barnard CN (1956) The genesis of intestinal atresia. Surg Forum 7:393–402

Benjamin B (1981) Endoscopy in esophageal atresia and tracheo-esophageal fistula. Ann Otol Rhinol 90:376–382

Berdon WE, Baker DH, Schulinger JN, Santulli TV (1979) Plain film detection of right aortic arch in infants with esophageal atresia and tracheoesophageal fistula. J Pediatr Surg 14:436–437

Berrocal T, Torres I, Guttierrez J et al (1999) Congenital anomalies of the upper gastrointestinal tract. Radiographics 19:855–872

Blumhagen JD, Rudd TG, Christie DL (1980) Gastroesophageal reflux in children: radionuclide gastroesophagography. AJR Am J Roentgenol 135:1001–1004

Boix-Ochoa J, Canals J (1976) Maturation of the lower esophagus. J Pediatr Surg 11:749–756

Bonnard A, Auber F, Fourcade L et al (2003) Vascular ring abnormalities: a retrospective study of 62 cases. J Pediatr Surg 38:539–543

Bremer JL (1944) Diverticula and duplications of the intestinal tract. Arch Pathol 38:132–140

Chacko J, Ford WDA, Furness ME (1998) Antenatal detection of hiatus hernia. Pediatr Surg Int 13:163–164

Chana J, Crabbe DC, Spitz L (1996) Familial hiatus hernia and gastro-oesophageal reflux. Eur J Pediatr Surg 6:175–176

Cheng W, Tam PKH (1999) Foreign-body ingestion in children: experience with 1,265 cases. J Pediatr Surg 34:1472–1476

Chetcuti P, Myers NA, Phelan PD et al (1989) Chest wall deformity in patients with repaired esophageal atresia. J Pediatr Surg 24:244–247

Collins P (1995) Chapter 3. Embryology and development. In: Banister LH, Berry MM, Collins P et al (eds) Gray's anatomy, 38th edn. Churchill Livingstone, Edinburgh, pp 91–342

Cumming WA (1975) Esophageal atresia and tracheoesophageal fistula. Radiol Clin North Am 13:277–295

Dantas RO, Dodds WJ, Massey BT, Kern MK (1989) The effect of high- vs low-density barium preparations on the quantitative features of swallowing. AJR Am J Roentgenol 153:1191–1195

DeMatteo C, Matovich D, Hjartarson A (2005) Comparison of clinical and video fluoroscopic evaluation of children with feeding and swallowing difficulties. Dev Med Child Neurol 47:149–157

Demirogullari B, Sonmez K, Turkyilmaz Z et al (2001) Colon interposition for esophageal stenosis in a patient with epidermolysis bullosa. J Pediatr Surg 36:1861–1863

Duncan M, Wong RKH (2003) Esophageal emergencies: things that will wake you from a sound sleep. Gastroenterol Clin N Am 32:1035–1052

Emblem R, Stringer MD, Hall CM, Spitz L (1993) Current results of surgery for achalasia of the cardia. Arch Dis Child 68:749–751

Engum SA, Grosfeld JL, West KW et al (1995) Analysis of morbidity and mortality in 227 cases of esophageal atresia and/or tracheoesophageal fistula over two decades. Arch Surg 130:502–508

Fasulakis S, Andronikou S (2003) Balloon dilatation in children for esophageal strictures other than those due to primary repair of oesophageal atresia, interposition or restrictive fundoplication. Pediatr Radiol 33:682–687

Fernbach SK, Glass RBJ (1988) The expanded spectrum of limb anomalies in the VATER association. Pediatr Radiol 18:215–220

Fernbach SK in: Gore RM, Levine MS, Laufer I (eds) (1994) Textbook of gastrointestinal radiology, 1st edn. WB Saunders Company. Philadelphia, pp 1380–1497

Fordham LA (2005) Imaging of the esophagus in children. Radiol Clin North Am 43:283–302

Gimenez A, Franquet T, Erasmus JJ et al (2002) Thoracic complications of esophageal disorders. Radiographics Spec No:S247–258

Gold B (2005) Asthma and gastroesophageal disease in children: exploring the relationship. J Peds 146:S13–S20

Gorenstein A, Cohen AJ, Cordova C et al (2001) Hiatal hernia in pediatric gastroesophageal reflux. J Pediatr Gastroenterol Nutr 33:554–557

Greil GF, Kramer U, Dammann F, Schick F et al (2005) Diagnosis of vascular rings and slings using an interleaved 3D double-slab FISP MR angiography technique. Pediatr Radiol 35:396–401

Griscom NT, Martin TR (1990) The trachea and esophagus after repair of esophageal atresia and distal fistula: computed tomographic observations. Pediatr Radiol 20:447–450

Harned RK, Strain JD, Hay TC, Douglas MR (1997) Esophageal foreign bodies: safety and efficacy of Foley catheter extraction of coins. AJR Am J Roentgenol 168:443–446

Hassal E (2005a) Decisions in diagnosing and managing chronic gastroesophageal reflux disease in children. J Pediatr 146:S3–S12

Hassal E (2005b) Outcomes of fundoplication: causes for concern, newer options. Arch Dis Child 90:1047–1052

Herman TE, Oser AB, McAlister WH (1991) Tubular communicating duplications of esophagus and stomach. Pediatr Radiol 21:494–496

Hernanz-Schulman M (2005) Vascular rings: a practical approach to imaging diagnosis. Pediatr Radiol 35:961–979

Hoeffel JC, Bigard MA, Merle M, Hoeffel C (1992) Epidermolysis bullosa: radiological patterns in childhood. Rofo 157: 427–428

Horn HM, Tidman MJ (2002) The clinical spectrum of dystrophic epidermolysis bullosa. Br J Dermatol 146:267–274

Hurwitz M, Bahar R, Ament ME et al (2000) Evaluation of the use of botulinum toxin in children with achalasia. J Pediatr Gastroenterol Nutr 30:509–514

Hussain SZ, Thomas R, Tolia V (2002) A review of achalasia in 33 children. Dig Dis Sci 47:2538–2543

Jadcherla SR, Duong HK, Hofmann C et al (2005) Characteristics of upper esophageal sphincter and oesophageal body during maturation in healthy human neonates compared with adults. Neurogastroenterol Motil 17:663–670

Kappelman MM, Dorst J, Haller JA et al (1969) H-type tracheo-esophageal fistula. Diagnostic and operative management. Am J Dis Child 118:568–575

Karasick S, Lev-Toaff AS (1995) Esophageal strictures: findings on barium radiographs. AJR Am J Roentgenol 165:561–565

Kern IB, Eisenberg M, Willis S (1989) Management of oesophageal stenosis in epidermolysis bullosa dystrophica. Arch Dis Child 64:551–556

Khelif K, De Laet MH, Chaouachi B et al (2003) Achalasia of the cardia in Allgrove's (Triple A) syndrome: histopathological study of 10 cases. Am J Surg Pathol 27:667–672

Koumanidou C, Vakaki M, Pitsoulakis G et al (2004) Sonographic measurement of the abdominal esophagus length in infancy: a diagnostic tool for gastroesophageal reflux. AJR Am J Roentgenol 183:801–807

Kovesi T, Rubin S (2004) Long term complications of esophageal atresia and/or tracheoesophageal fistula. Chest 126:915–925

Kramer SS (1989) Radiologic examination of the swallowing impaired child. Dysphagia 3:117–125

Kuhns LR, Poznanski AK (1972) A device for control of esophageal tubes in infants. Radiology 102:438–439

Leithiser RE, Capitanio MA, Macpherson RI, Wood BP (1985) "Communicating" bronchopulmonary foregut malformations. AJR Am J Roentgenol 146:227–231

Levine MS, Rubesin SE (2005) Diseases of the esophagus: diagnosis with esophagography. Radiology 237:414–427

Lifschitz CH (2001) Feeding problems in infants and children. Curr Treat Options Gastroenterol 4:451–457

Martinez L, Rivas S, Hernandez F et al (2003) Aggressive conservative treatment of esophageal perforations in children. J Pediatr Surg 38:685–689

Mayberry JB, Mayell MJ (1998) Epidemiological study of achalasia in children. Gut 29:90–93

McGuirt WF (2003) Gastroesophageal reflux and the upper airway. Pediatr Clin N Am 50:487–502

Mercado-Deane MG, Burton EM, Harlow SA et al (2001) Swallowing dysfunction in infants less than 1 year of age. Pediatr Radiol 31:423–428

Meschan I (1975) An atlas of anatomy basic to radiology. WB Saunders Company, Philadelphia

Michel JL, Revillon Y, Salakos C et al (1997) Successful bronchotracheal reconstruction in esophageal bronchus: two case reports. J Pediatr Surg 32:739–742

Miller CK, Willging JP (2003) Advances in the evaluation and management of pediatric dysphagia. Curr Opin Otolaryngol Head Neck Surg 11:442–446

Murphy SG, Yazbeck S, Russo P (1995) Isolated congenital esophageal stenosis. J Pediatr Surg 30:1238–1241

Nelson SP, Chen EH, Syniar GM, Christoffel KK (1997) Prevalence of symptoms of gastroesophageal reflux during infancy: a pediatric practice-based survey. Arch Pediatr Adolesc Med 151:569–572

Newman B, Bender TM (1997) Esophageal atresia/tracheoesophageal fistula and associated congenital esophageal stenosis. Pediatr Radiol 27:530–534

Newman LA, Keckley C, Petersen MC, Hammer A (2001) Swallowing function and medical diagnoses in infants suspected of dysphagia. Pediatrics 108:E106

Noel RJ, Rothenberg ME (2005) Eosinophilic esophagitis. Curr Opin Pediatr 17:690–694

Poznanski A (1969) A simple device for administering barium to infants. Radiology 93:1106

Quan L, Smith WL (1973) The VATER association: vertebral defects, anal atresia, T-E fistula with esophageal atresia, radial and renal dysplasia: a spectrum of associated defects. J Pediatr 82:104–107

Rudolph CD, Mazur LJ, Liptak GS et al (2001) Guidelines for evaluation and treatment of gastroesophageal reflux in infants and children: recommendations of the North American Society for Pediatric Gastroenterology and Nutrition. J Pediatr Gastroenterol Nutr 32:S1–S32

Sadler TW (2000) Langman's medical embryology, 8th edn. Lippincott Williams and Wilkins, Baltimore, MD

Sant'Anna AM, Rolland S, Fournet JC et al (2004) Eosinophilic esophagitis in children: symptoms, histology, and pH probe results. J Pediatr Gastroenterol Nutr 39:373–377

Schlesinger AE, Parker BP in: Kuhn JP, Slovis TL, Haller JO (eds) (2004) Caffey's pediatric diagnostic imaging, 10th edn. Mosby, Philadelphia, pp 1539–1583

Schunk JE, Harrison MA, Corneli HM, Nixon GW (1994) Fluoroscopic Foley catheter removal of esophageal foreign bodies in children: experience with 415 episodes. Pediatrics 94:709–714

Seibert JJ, Byrne WJ, Euler A, et a. (1983) Gastroesophageal reflux – the acid test: scintigraphy or the pH probe? AJR Am J Roentgenol 140:1087–1090

Sharieff GQ, Brousseau TJ, Bradshaw JA, Shad JA (2003) Acute esophageal coin ingestion: is immediate removal necessary? Pediatr Radiol 33:859–863

Shaw-Smith CJ (2005) Oesophageal atresia, tracheo-esophageal fistula and the VACTERL association: review of genetics and epidemiology. J Med Genet 43:545-554 [Epub ahead of print]

Singleton EB, King BA (1971) Localized lesions of the stomach in children. Semin Roentgenol 6:220–234

Sor S, Levine MS, Kowalski TZ et al (1995) Giant ulcers of the esophagus in patients with human immunodeficiency virus: clinical, radiographic, and pathologic findings. Radiology 194:447–451

Srikanth MS, Ford EG, Stanley P, Mahour GH (1992) Communicating bronchopulmonary foregut malformations: classification and embryogenesis. J Pediatr Surg 27:732–736

Vaezi MF, Richter JE (1999) Current therapies for achalasia: comparison and efficacy. J Clin Gastroenterol 27:21–35

Vane DW, Cosby C, West K, Grosfeld JL (1988) Late results following esophagomyotomy in children with achalasia. J Pediatr Surg 23:515–519

Vasudevan SA, Kerendi F, Lee H, Ricketts RR (2002) Management of congenital esophageal stenosis. J Pediatr Surg 37:1024–1026

Vazquez JL, Buonomo C (1999) Feeding difficulties in the first days of life: findings on upper gastrointestinal series and the role of videofluoroscopic swallowing study. Pediatr Radiol 29:894–896

Walor D, Berdon W, Anderson N et al (2005) Gaseous distention of the cervical esophagus with nasal CPAP: mimicker of pharyngeal perforation and esophageal atresia. Pediatr Radiol 35:1196–1198

Wang JK, Chang MH, Li YW et al (1993) Association of hiatus hernia with asplenia syndrome. Eur J Pediatr 142:418–420

Webb WA (1995) Management of foreign bodies of the upper gastrointestinal tract: update. Gastrointest Endosc 41:39–51

Westra SJ, Wolf BH, Staalman CR (1990) Ultrasound diagnosis of gastroesophageal reflux and hiatal hernia in infants and young children. J Clin Ultrasound 18:477–485

Woods RK, Sharp RJ, Holcomb GW 3rd et al (2001) Vascular anomalies and tracheoesophageal compression: a single institution's 25-year experience. Ann Thorac Surg 72:438–439

Yardeni D, Yardeni H, Coran AG, Golladay ES (2004) Severe esophageal damage due to button battery ingestion: can it be prevented? Pediatr Surg Int 20:496–501