The Esophagus

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Abstract

The esophagus is a dynamic conduit, which connects the oral cavity to the stomach. Esophageal secretions and antegrade peristalsis enable the passage of solids and liquids toward the stomach. Conversely, the esophagus enables prompt expulsion of gastrointestinal content during vomiting and passively with gastric reflux. The pediatric esophagus is subject to a number of congenital and acquired disorders. In this chapter, we review the embryology, applied anatomy, and diagnostic imaging appearances related to the normal pediatric esophagus and in infants and children with esophageal disease.

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1 Anatomy and Development of the Esophagus

1.1 Embryology

The esophagus and trachea are derived from a common tubular structure, which differentiates into the esophagus posteriorly and laryngotracheal groove anteriorly (Fig. 1). The respiratory diverticulum forms on the ventral wall of the primitive foregut at the border of the pharyngeal gut. The esophagus can be recognized as a structure distinct from the pharynx and stomach by the third gestational week at a fetal crown-rump-length (CRL) of 2.5 mm (Takubo 2009). Regional differentiation of the esophagus and stomach from the remainder of the gastrointestinal tract is specified by SOX2 genes (Sadler 2012). The mesodermal

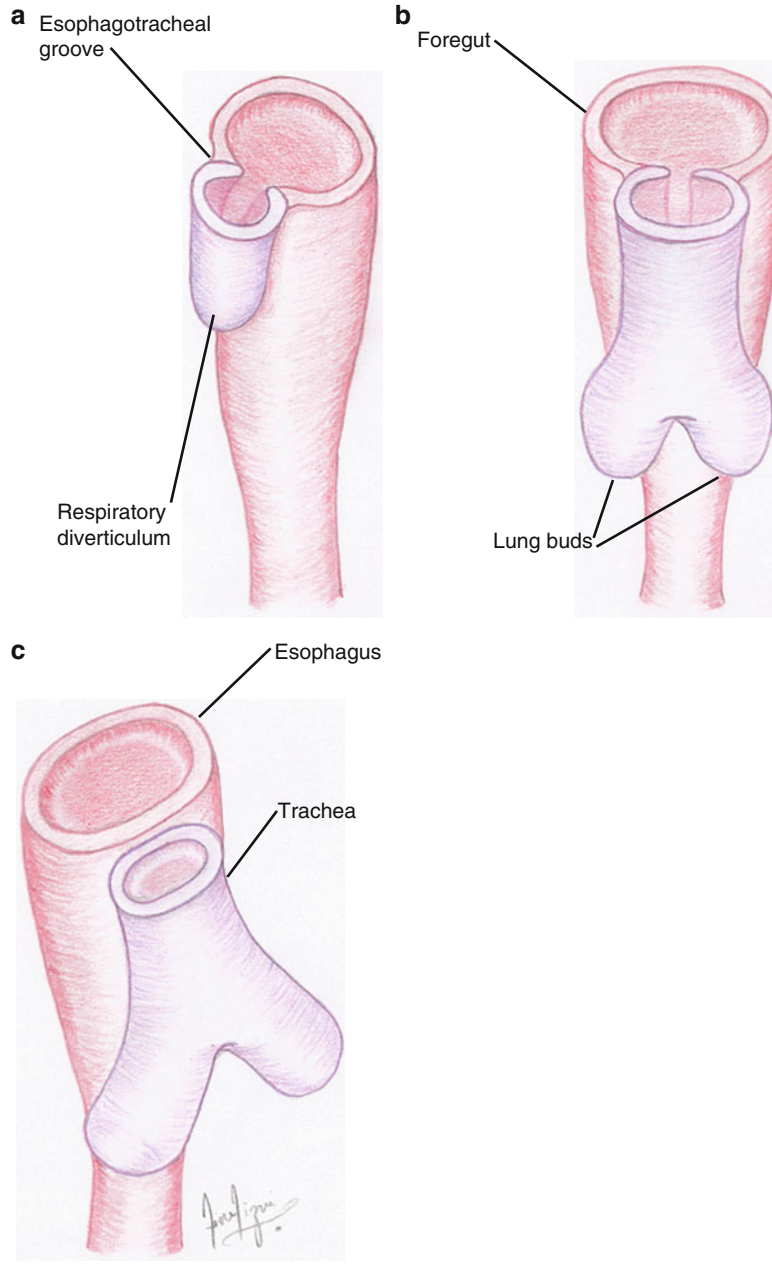

Fig. 1 (**a**–**c**) Embryology of the esophagus. (**a**) The respiratory diverticulum arises from the ventral wall of the foregut at 4 weeks (**b**–**c**). The laryngotracheal groove partitions the trachea from the primitive foregut, the latter eventually differentiates into the esophagus. Image courtesy of Dr. Aisha Rizvi MBBS, Doha, Qatar

esophagotracheal septum undergoes progressive bilateral evagination partitioning the dorsal esophagus from the ventral respiratory epithelium. Most of the commonest congenital esophageal abnormalities relate to failure of this partitioning process. The esophagus is initially short and only attains normal relative length by the 7th week. Failure to elongate satisfactorily pulls the stomach cranially through the esophageal hiatus resulting in a congenital hiatus hernia.

1.2 Imaging Anatomy

The esophagus is a muscular conduit. It extends from the pharynx just left of midline at the level of the C6 vertebral body and inferior cricoid cartilage. Along the length of the neck, the esophagus lies posterior to the trachea and anterior to the cervical prevertebral muscles. The recurrent laryngeal nerves pass between the esophagus and trachea. Laterally it is bound by each carotid sheath (containing the vagus nerve, internal jugular vein, and common carotid artery) and the lateral lobes of the thyroid.

In the thorax the esophagus courses inferiorly and slightly obliquely to the left. It lies predominantly within the posterior mediastinum adjacent to the thoracic prevertebral muscles and anterior to the thoracic duct, posterior intercostal arteries, and azygos veins. The anterior surface of the esophagus abuts the aortic arch, left mainstem bronchus at around the T5 vertebral level, and left atrial parietal pericardium. The esophagus lies between the layers of the parietal pleura. The azygoesophageal recess (AER) is an anatomical space lying anterior to the spine and posterolateral to the lower thoracic esophagus. The AER extends from the level of the anterior turn of the azygos vein at T4 and inferiorly to the level of the aortic hiatus (Gibbs et al. 2007). The AER is formed by the interface of the azygos vein, lower thoracic esophagus, and posteromedial right lung lower lobe pleura. Distortion of the AER contour can be detected on a radiograph or cross-sectional imaging and may imply pathology such a hiatus hernia or bronchopulmonary foregut malformation.

The thoracic duct lies to the right and posterior aspect of the esophagus at T5 and extends cranially to the neck. The esophagus enters the abdomen through the diaphragmatic esophageal hiatus formed by a muscular sling from the right hemidiaphragmatic crus just to the left of the midline at T10. The intra-abdominal portion of the esophagus lies anterior to the left crus of the diaphragm and posterior to the left hepatic lobe. The esophagus terminates at the gastric cardia at T11. The distal end of the esophagus forms into a slightly dilated vestibule just above the gastroesophageal junction (GEJ). The upper limit of the vestibule is at the A-ring, the lower limit at the B-ring (Fig. 2). The A- and B-rings are usually only visualized in the presence of a small sliding hiatus hernia. The Z-line refers to the junction of the esophageal and gastric mucosa and may occasionally be seen as a subtle line on a double contrast barium study.

The esophagus is composed mainly of striated muscle in its upper third, smooth muscle in the distal third, and a mixture of the two in its

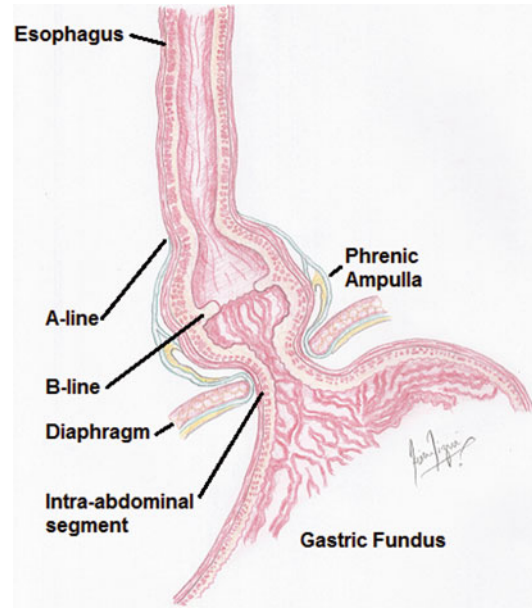

Fig. 2 Esophageal rings. Line diagram showing the position of the muscular A-ring at the junction between the phrenic ampulla and tubular esophagus, and mucosal B-line demarcating the squamocolumnar junction. Image courtesy of Dr. Aisha Rizvi MBBS, Doha, Qatar

mid-portion. The upper esophageal sphincter (UES) and normal antegrade peristaltic activity are developed by 33 weeks gestation (Jadcherla et al. 2005). The muscularis propria is comprised of smooth muscle and its inner circular layer forms in the 6th week. The outer longitudinal layer is complete by the 10th week. Stratified muscle develops in the fourth month. The muscularis mucosae develops cranially from the distal esophagus in the fourth month and is complete by the 7th month. The circular muscle layer provides the sequential antegrade peristaltic contractions toward the stomach. It is continuous with the hypopharyngeal inferior constrictor muscle. The circular muscle layer courses transversely in the cranial and caudal regions of the esophagus and obliquely within the body of the esophagus and is continuous with the intrinsic component of the lower esophageal sphincter (see below). The UES is located between the pharynx and the cervical esophagus. It is comprised of the hyoid bone, the posterior aspects of the cricoid and thyroid cartilage, and three muscles: thyropharyngeus, inferior esophageal constrictors, and cricopharyngeus. The cricopharyngeus muscle is transversely oriented, while thyropharyngeus muscle is obliquely oriented. The region between these two muscles is the Killian triangle (dehiscence), from which a Zenker diverticulum may form; the latter almost exclusively develops in late adulthood. The cricopharyngeus muscle lies at C5–C6 and is normally relaxed on swallowing, hence not normally visualized on contrast esophagram but may be seen as a posterior horizontal impression in a small number of otherwise asymptomatic patients. Cricopharyngeal dysfunction however may cause dysphagia. The lower esophageal sphincter (LES) is located where the esophagus merges with the stomach. It is comprised of intrinsic and extrinsic components. The intrinsic component consists of esophageal muscle fibers under neurohormonal influence. The extrinsic component consists of the esophageal hiatus in the crural diaphragm and serves as an external sphincter. Malfunction of either component may result in gastroesophageal reflux (GER). The manometric location of the LES differs from the endoscopic location.

The arterial blood supply of the esophagus is extensive comprising of a segmental network of anastomotic vessels. Branches of the inferior thyroid artery supply the upper esophageal sphincter (UES) and cervical esophagus. Paired aortic esophageal arteries or terminal bronchial arteries supply the thoracic esophagus. The distal esophagus and LES are supplied by the left gastric artery and branches of the left phrenic artery. The venous drainage of the esophagus is derived from an extensive submucosal plexus that drains into the superior vena cava via the azygos system. Collaterals of the left gastric vein, a branch of the portal vein, receive venous drainage specifically from the mid-esophagus. Submucosal connections between the portal and systemic venous systems in the distal esophagus enable esophageal varices to form in portal hypertension resulting in submucosal varices. These are sources of major upper gastrointestinal hemorrhage in conditions such as liver cirrhosis (Kuo and Urma 2006). Esophageal lymphatic egress is derived from three interconnected drainage systems. The proximal third drains into the thoracic duct via deep cervical lymph nodes. The middle third drains into the superior and posterior mediastinal nodes. The distal third follows the venous drainage along the course of the left gastric artery to the gastric and celiac lymph nodes. The innervation of the esophagus includes the Meissner plexus, which provides vagal secretomotor parasympathetic innervation of the submucosa, and the Auerbach (myenteric) plexus which provides spinal motor innervation of the muscularis propria.

2 Imaging Techniques and Indications

2.1 Radiography

The non-distended esophagus is not visualized on plain radiography and can be difficult to assess on cross-sectional imaging. The position of the esophagus may be inferred on radiographs by the presence of intraluminal air. An air-filled esophagus typically results from aerophagia, during

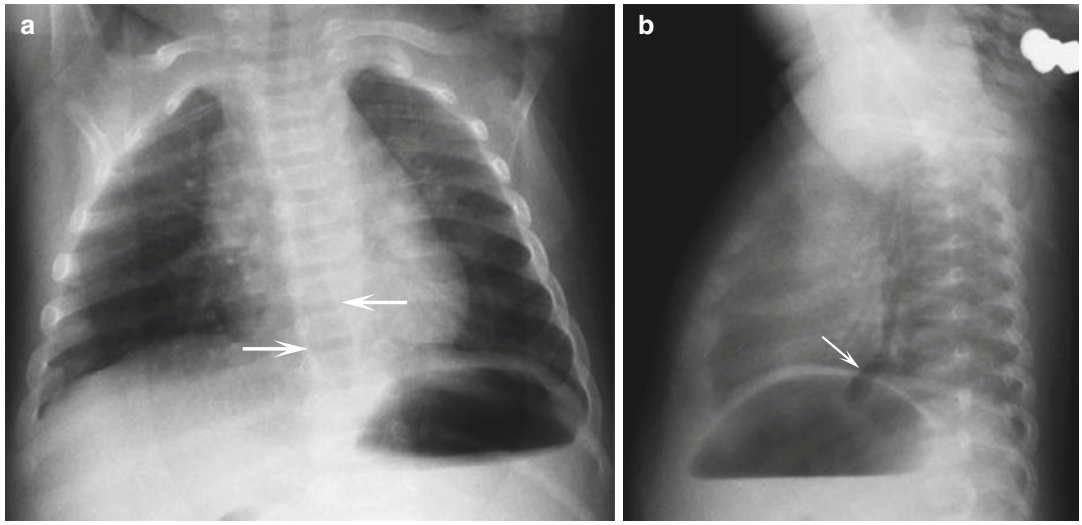

Fig. 3 (**a**, **b**) Air outlining a normal esophagus. (**a**) AP and (**b**) lateral chest radiographs of a crying infant with an air-filled esophagus (*arrows*) secondary to aerophagia

crying, when large amounts of air are swallowed (Fig. 3). In neonates continuous positive pressure may result in marked gaseous distension of the esophagus and stomach requiring decompression via an orogastric tube. Segmental gaseous distension of the esophagus is seen in an atretic esophageal pouch or proximal to an esophageal stricture. Tracheoesophageal fistula is another cause of an air-filled esophagus. An air-fluid level may be visualized on an erect chest radiograph in esophageal stricture or achalasia. Radiographs are important in evaluating the course and termination of orogastric and nasogastric tubes (Figs. 4 and 5). The position of an intact esophagus may be inferred from the course of an indwelling enteric tube. Aberrant course raises the possibility of esophageal perforation (Fig. 6) or other pathology distorting the esophagus.

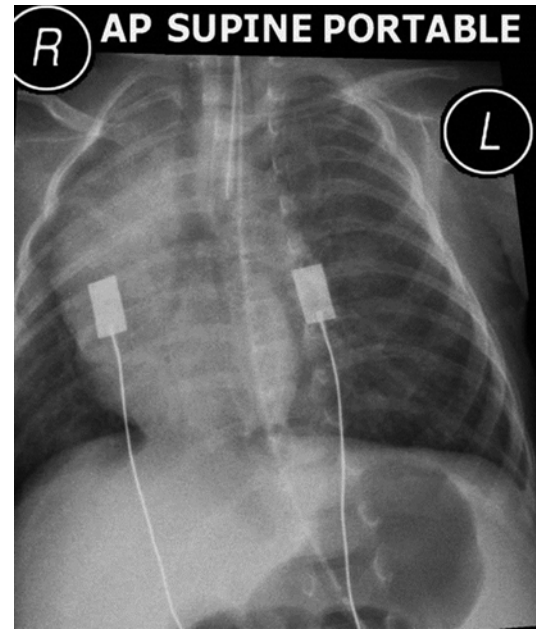

Fig. 4 Malposition of endotracheal tube. Esophageal malposition of an endotracheal tube. Oblique frontal chest radiograph confirming inadvertent intubation of the esophagus with an endotracheal tube posterior to the airway

2.2 Fluoroscopy

Fluoroscopy remains the key imaging modality to assess the pediatric and adolescent esophagus. The contrast esophagram (or barium swallow) is the radiological imaging test of choice to evaluate the anatomy, dysphagia, and dysmotility. It is preferred in the initial work-up and follow-up of most surgical esophageal disorders. The esophagram provides comprehensive intraluminal anatomical information from which mural and extraluminal pathology may also be inferred. It

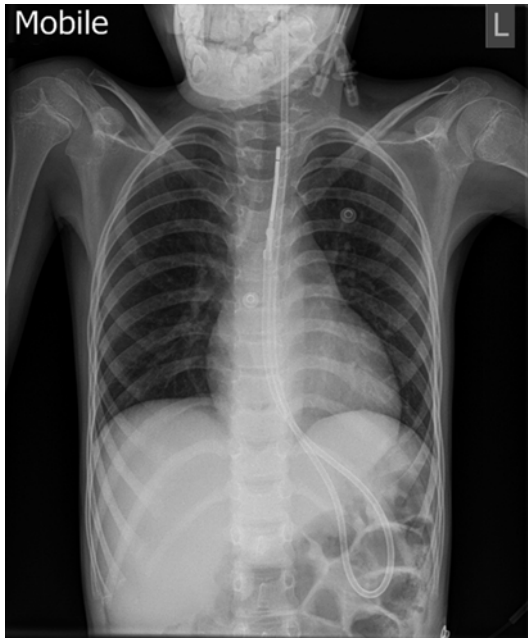

Fig. 5 Esophageal termination of a nasogastric tube. Suboptimal enteric tube position with the tube coiling back on itself in the gastric cardia and terminating in the esophagus. This is not safe for feeding and should be removed and replaced

also yields detailed functional information regarding transit time and patency of the gastroesophageal sphincters. The esophagram can be coupled with an upper gastrointestinal (UGI) series (Fig. 7) to assess the stomach and proximal small bowel in order to detect GER and exclude structural abnormalities causing vomiting. Ideally the stomach and proximal small bowel should be imaged as part of the initial esophagram in a child to document gastric emptying and duodenojejunal flexure position. Follow-up studies should include the proximal stomach. The main indications for performing an esophagram in children are summarized in Table 1. The choice of contrast agent used is at the discretion of the radiologist who should be knowledgeable of any patient allergies and the specific clinical indication. Barium sulfate yields the highest-quality single and double contrast studies. Assessment of fine mucosal detail is rarely required in children and single contrast studies usually suffice. In sick neonates or where there is suspected contrast leakage, low-osmolar water-soluble contrast medium is preferable. Gastrografin by virtue of

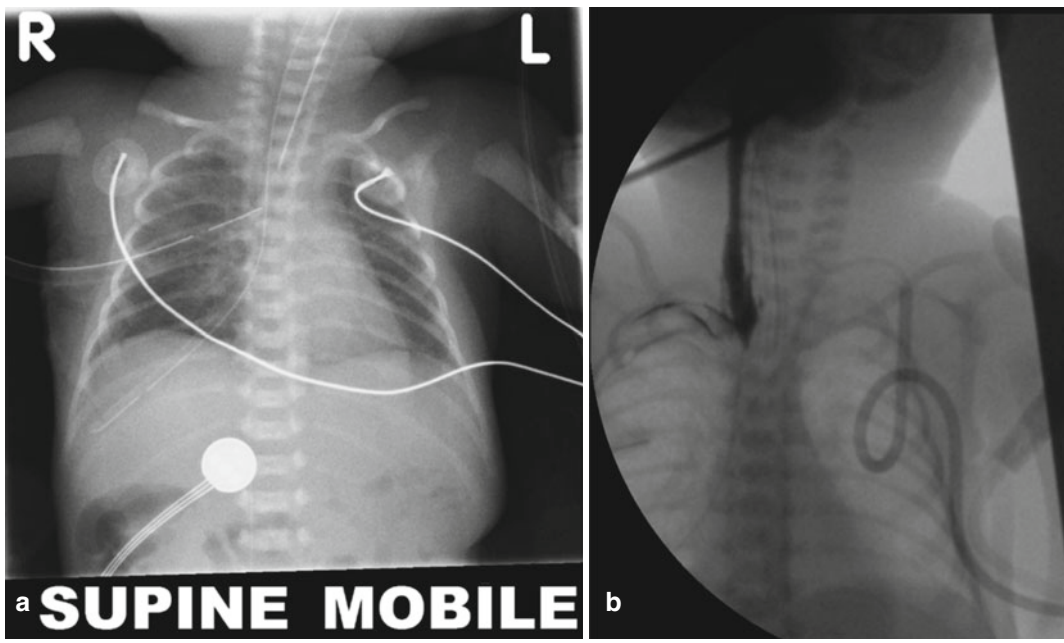

Fig. 6 (**a**, **b**) Iatrogenic esophageal perforation. (**a**) Frontal chest radiograph showing abnormal course of the orogastric tube terminating in the right upper abdominal quadrant. (**b**) Water-soluble contrast series showing a contained extrapleural contrast leak

its high osmolality is to be avoided in all children and patients with neurological impairment, as pulmonary aspiration has been associated with fatal pulmonary edema. Gastromiro® is favored in Europe and gastrografin is not routinely used in UGI series. Oral intake of contrast medium is to be encouraged and can be made more palatable using a range of flavorings. Caution is required when opacifying the esophagus using an enteric tube. It may be difficult to control opacification with the risk of inadvertent pulmonary aspiration leading to transient hypoxia or even respiratory arrest in the labile child (Fig. 8). One scenario where such a technique is employed is the post-operative assessment of tracheoesophageal fistula (see Sect. 4.1). The infant is placed prone and lateral shoot-through or biplane fluoroscopy is performed at a high frame rate. The aim of the

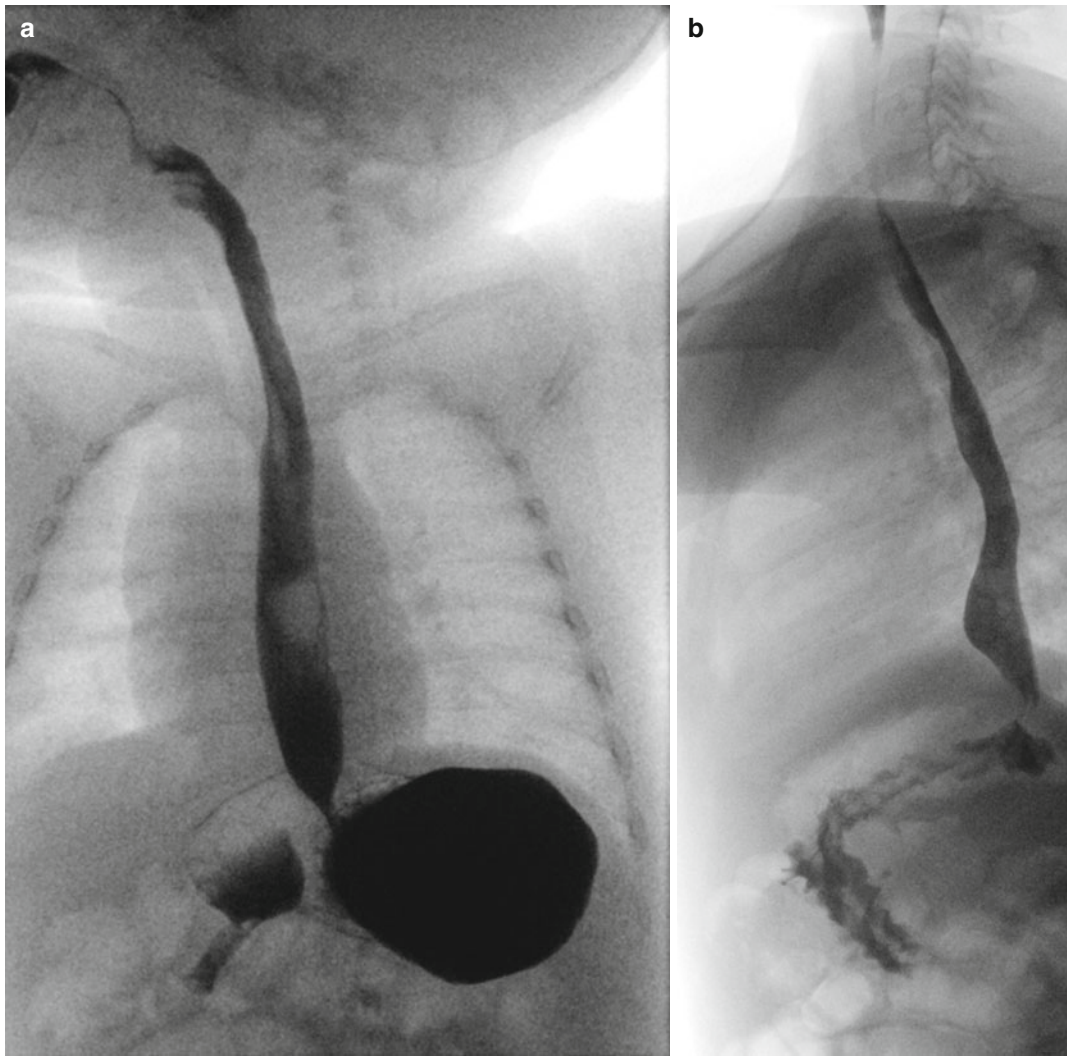

Fig. 7 (**a**–**d**) Normal barium esophagram in an infant. (**a**) Oblique anteroposterior and (**b**) lateral barium esophagrams, the lateral esophagram shows normal anterior indentation of the esophagus by the aortic arch, left main bronchus, and left atrium. (**c**) AP esophagram in a 9-month-old child showing normal parallel orientation of the mucosal folds in the empty esophagus (*arrow*). The esophageal mucosal folds should be less than five in number with a maximal thickness of 3 mm. (**d**) Spurious mobile lucent gaseous filling defects due to excessive crying, these promptly cleared

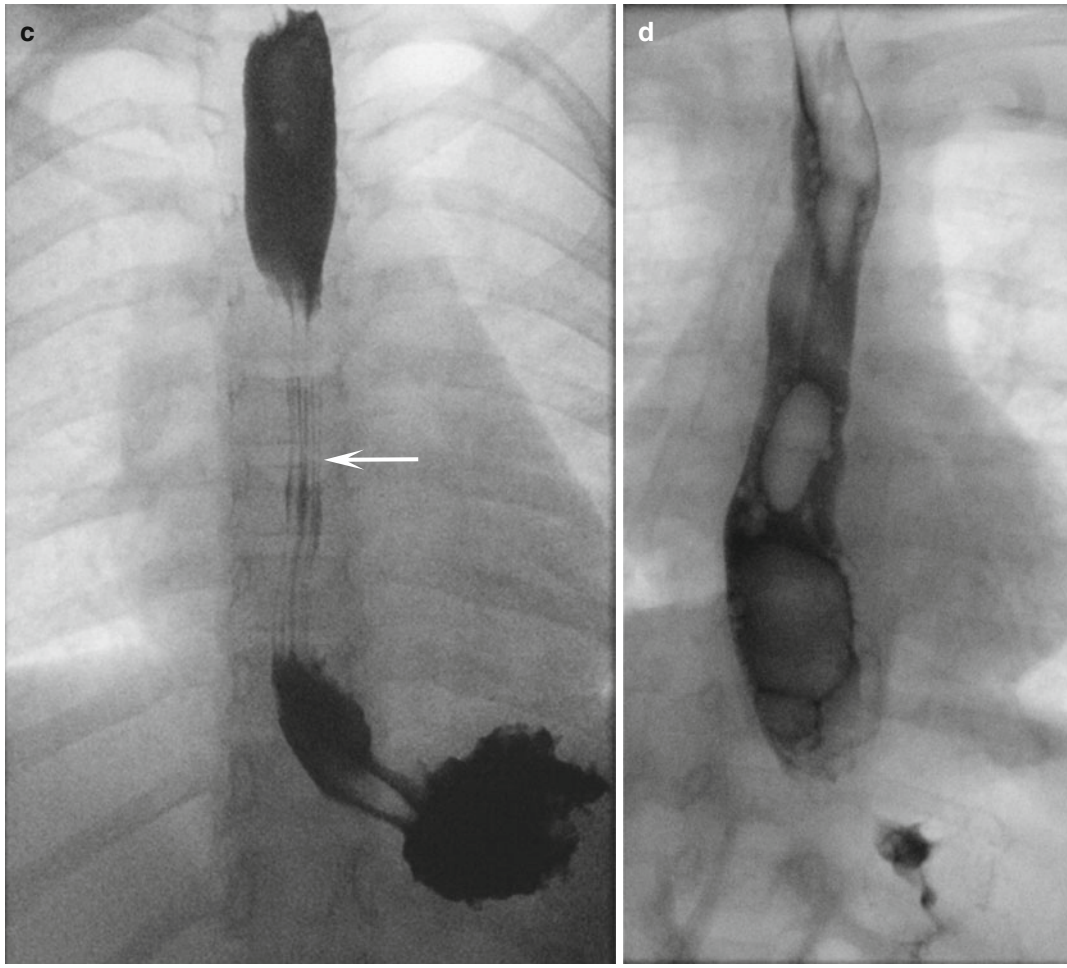

Fig. 7 (continued)

Table 1 Summary of the main indications for an esophagram

Dysphagia
Odynophagia
Dysmotility disorders
Recurrent pneumonia or chronic tracheobronchial inflammation (aspiration)
Strictures
Extrinsic compression (vascular or non-vascular)
Esophageal obstruction
Postsurgical evaluation
Esophagitis
Gastroesophageal reflux
Tracheoesophageal fistula
Chest pain of non-cardiac origin
Varices
Neoplasm

esophagram is to adequately image the distended esophagus in two planes without aspiration. The cervical esophagus is assessed in the true antero-posterior (AP) and lateral planes. The AP and lateral planes should be used for the thoracic esophagus with obliquity as required to avoid overlap from the spine. In most cases normal feeding volumes are most appropriate. If the child refuses to drink adequate volumes of contrast, then the contrast volume as a last resort may be augmented with a more palatable drink such as juice or milk. The sated child may refuse to drink and ingestion of a recent meal may result in spurious intraluminal findings from retained ingested matter; hence a pre-procedural fast is essential. Aerophagia can mimic intraluminal

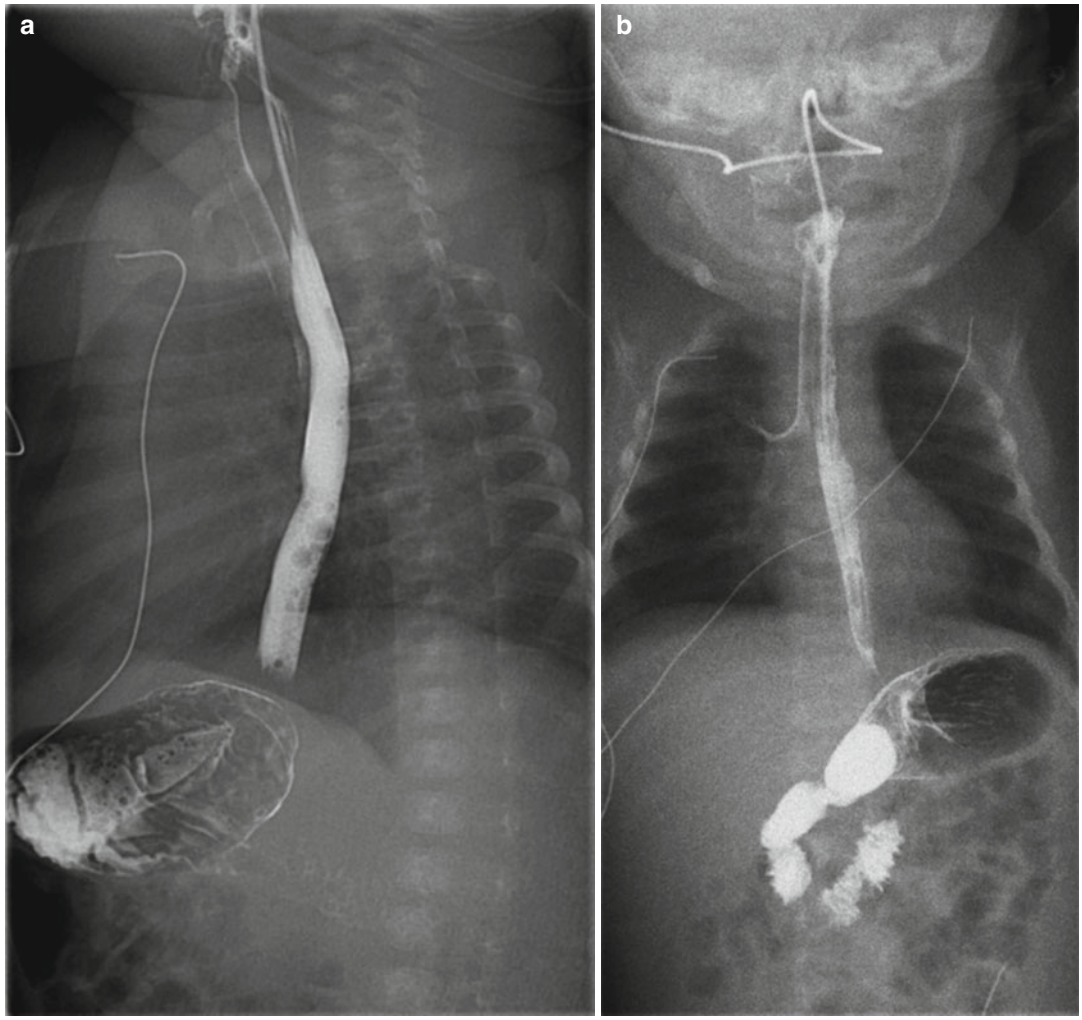

Fig. 8 (**a**, **b**) Overflow aspiration during tube-esophagram study. (**a**) Lateral and (**b**) AP esophagography show high enteric tube position and overdistension of the esophagus with unintentional contrast overflow into the airway

filling defects (Fig. 7d). Fasting times depend on the child's age and should not exceed the child's routine feeding intervals. Age-appropriate fasting times are in the order of approximately 2–3 h for neonates and young infants, 4 h for older infants and children, and at least 6–8 h for adolescents (ACR 2014). Fasting children are best scheduled for early morning examinations to minimize patient distress and parental anxiety. Infants and younger children are examined recumbent and may require immobilization to reduce motion artifact and minimize fluoroscopy time. Older children and adolescents may prefer swallowing barium in the erect position. Esophageal transit is aided by gravity. A double contrast study may be performed in older children using barium sulfate and Carbex® and completed with the ingestion of a small absorbable barium tablet. The latter may be retained in subtle disorders of esophageal dysmotility.

As with all fluoroscopic imaging, radiation exposures must be as low as reasonably achievable using pediatric-specific protocols, employing low-dose, pulsed and cine-fluoroscopy with grabbed images and cine-loop capture. Fine mucosal detail and depiction of

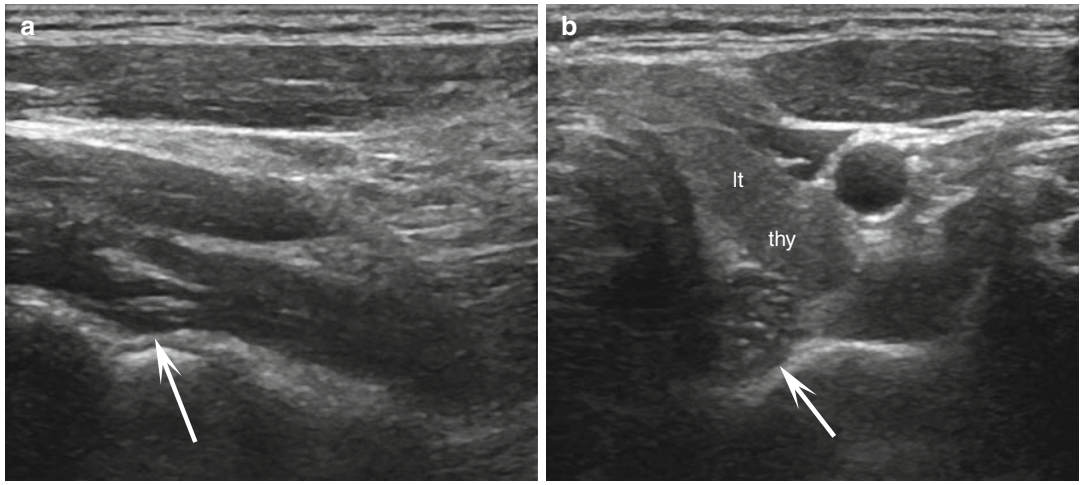

Fig. 9 (**a**, **b**) Ultrasound of the cervical esophagus. (**a**) Longitudinal and (**b**) transverse views of the cervical esophagus (*arrow*) obtained using the left thyroid lobe (lt thy) as an acoustic window

subtle contrast leakage or fistulae may require higher dose exposures. The assessment of dysmotility is typically performed as part of a multidisciplinary team with a speech and language therapist attending the study. The child is assessed with different age-appropriate consistencies of food and liquid ranging from barium-coated solids (fruit or small cookie pieces), thickened puree or yogurt, and thin liquids (see Sect. 3).

2.3 Ultrasound

Ultrasound (US) is not an organ-specific imaging technique. It is, however, safe and widely available. US to date has not been widely adopted to assess the esophagus. Portions of the esophagus can be noninvasively imaged using high-frequency high-resolution linear transducers. The esophagus has five layers best appreciated using endoscopic US. Novel applications of conventional US include the assessment of swallowing and lower esophageal sphincter competency. US can be used to dynamically assess the tongue, oral cavity, hyoid, and larynx during swallowing using the midline anterior neck as an acoustic window (Miller and Kang 2007). The cervical esophagus can be assessed using the thyroid gland as an acoustic window (Fig. 9). The intra-abdominal portion of the esophagus may be evaluated through the left lobe of the liver (Fig. 10) to dynamically depict gastroesophageal reflux following a feed (Koumanidou et al. 2004). The esophagus itself serves as an important acoustic window for the echocardiographer using transesophageal echocardiography.

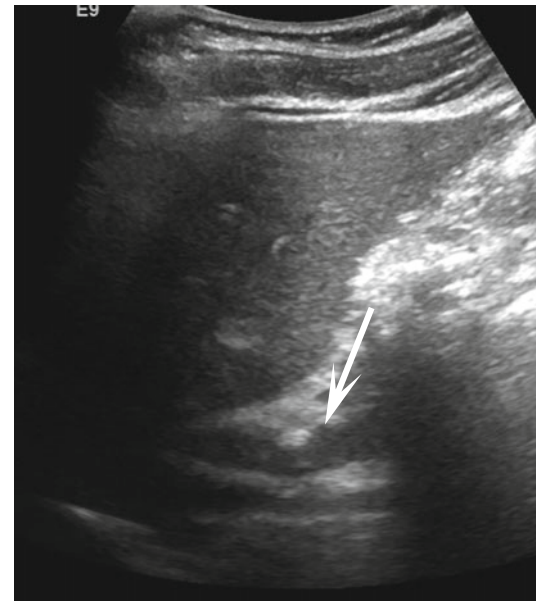

Fig. 10 Ultrasound of the intra-abdominal esophagus. Longitudinal US image of the esophagus (*arrow*) obtained using the left lobe of the liver as an acoustic window

2.4 Computed Tomography

The esophagus is not primarily assessed using computed tomography (CT) in children. One specific exception is suspected extrinsic esophageal compression by a vascular ring or pulmonary arterial sling (see Sect. 4.6). Modern multi-detector CT (MDCT) enables rapid image acquisition with reduced sedation requirements. Imaging can be performed with free-breathing. 2–3 cc/kg of intravenous low-osmolar iodinated contrast medium is used; oral contrast medium is not required. Acquisition timing may be optimized using bolus tracking and threshold triggering. Weight-based exposure parameters should be used to ensure achieving as-low-as-reasonably possible radiation doses. MDCT can dynamically assess airway caliber. Two-dimensional (2D) multiplanar reformatted, maximum-intensity, minimum-intensity projection, and three-dimensional (3D) imaging reconstructions can be generated to improve diagnostic yield.

2.5 Magnetic Resonance Imaging

Magnetic resonance imaging (MRI) is not a first-line imaging strategy for the pediatric esophagus. The esophagus is subject to considerable cardiac and respiratory motion, and EKG-gated and respiratory-gated imaging techniques are required to optimally demonstrate the esophagus. MRI is an alternative to MDCT for assessing vascular rings but is more time-consuming and frequently requires sedation or general anesthesia. MRI can be useful for assessing esophageal involvement from (para)spinal pathology such as tumor. MR image post-processing uses techniques similar to those used for MDCT.

2.6 Scintigraphic Studies

In children scintigraphic studies may be used to (i) verify suspected gastroesophageal reflux or pulmonary aspiration, (ii) assess esophageal transit, and (iii) quantify the rate of gastric emptying of liquid meals. In infants and children, a gastroesophageal reflux study is typically combined with a liquid phase gastric emptying study ("milk scan"). Techniques are not well standardized across institutions. Practice parameter guidelines for performing gastrointestinal scintigraphy have been jointly issued by American College of Radiology, Society for Nuclear Medicine, and Society for Pediatric Radiology (ACR–SNM–SPR 2014) for use in children. A pre-procedural fast of approximately 4 h is required prior to administering a liquid meal consisting of a weight-based concentration of technetium-99 m (Tc-99 m) sulfur colloid and either milk, formula, or juice. The feed can be administered orally, by nasogastric tube, or via a percutaneous gastrostomy tube. Dynamic imaging of the chest and abdomen is obtained in the supine position for 60 min. The number of reflux events during the study, duration, and proximal extent of reflux can be reported (Fig. 11). Pulmonary aspiration is confirmed by activity within the lungs on delayed imaging. Gastric emptying is assessed at 1, 2, and 3 h after completion of feeding. Follow-up studies can be used to assess response to medical therapy. Contrast studies coupled with manometry in most cases have obviated need for dynamic supine esophageal scintigraphic transit studies; however, these can be performed before or after a combined milk scan.

3 Swallowing Disorders

3.1 Normal Swallowing and Esophageal Transit

Swallowing is a complex dynamic sequential process that consists of both voluntary and involuntary components. The voluntary phases include preparatory sucking and oral bolus formation. In the oral phase coordinated movement of the mandible along with elevation of the tongue toward the hard palate propels the ingested bolus posteriorly toward the oropharynx. The involuntary components comprise the pharyngeal and esophageal phases. In the pharyngeal phase elevation the soft palate by the arriving bolus causes the palatine constrictor muscles to contract and the bolus passes into the oropharynx. Simultaneously

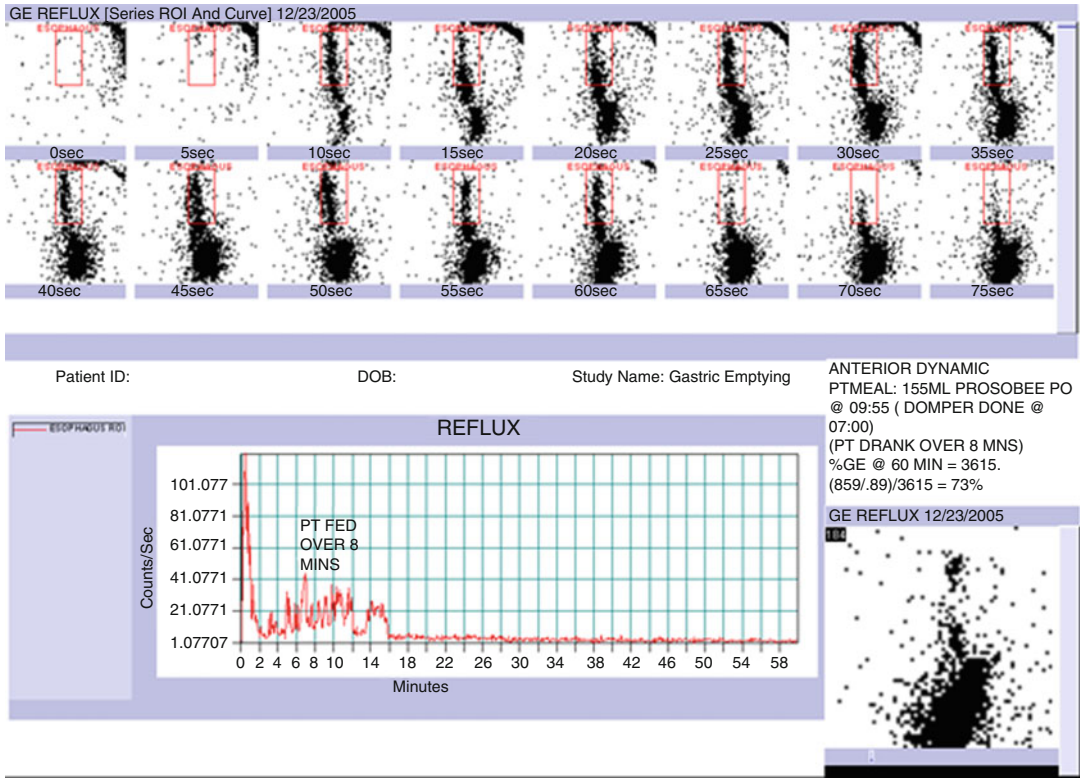

Fig. 11 Tc99m radionuclide "milk" scan. Serial imaging demonstrates gastroesophageal reflux to the upper esophagus but no pulmonary aspiration on delayed imaging at 4 h

the velopharyngeal portal closes to prevent nasal escape of the bolus. The oropharyngeal constrictor muscles generate a peristaltic wave toward the larynx, closing the vocal cords, the arytenoids descend narrowing the laryngeal vestibule while depressing the epiglottis, clearing any laryngeal penetration and preventing pulmonary aspiration. Respiratory ventilation ceases. The bolus passes laterally around the epiglottis toward the piriform sinuses and then through the upper esophageal sphincter (UES). The UES comprises of the inferior pharyngeal constrictor muscles, cricopharyngeus muscle, and proximal cervical esophagus. The UES is normally tensioned at rest. Preceding bolus arrival, the cricopharyngeus muscle relaxes. The suprahyoid muscles and thyrohyoid muscles contract, the hyolaryngeal complex moves anteriorly opening the UES (Matsuo and Palmer 2008). As the bolus enters the UES, primary peristalsis consists of an initial rapid primary wave of relaxation to accommodate a bolus and a slower secondary wave of contraction that propels it toward the lower esophageal sphincter (LES). Gravity aids peristalsis in the upright position. The LES at rest prevents gastric reflux but relaxes during a swallow and allows bolus passage into the stomach. Secondary peristalsis ensures any residual bolus passes into the stomach and is initiated locally in the esophagus by luminal distension by the retained bolus. Tertiary contractions are abnormal non-propulsive contractions and signify esophageal dysmotility (Fig. 12).

3.2 Disorders of Swallowing

The prevalence of feeding disorders at any point in a developing child varies but is estimated to be between 25 and 40 % (Lefton-Greif and Arvedson 2007). Incidence levels are much higher in

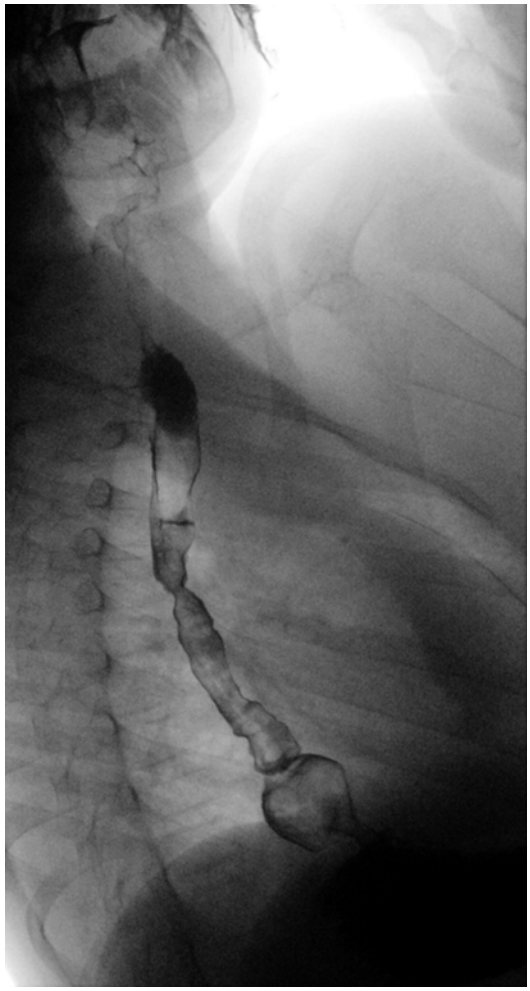

Fig. 12 Esophageal dysmotility. Barium esophagram showing a corkscrew appearance to the esophagus in a child with diffuse esophageal spasm and tertiary contractions

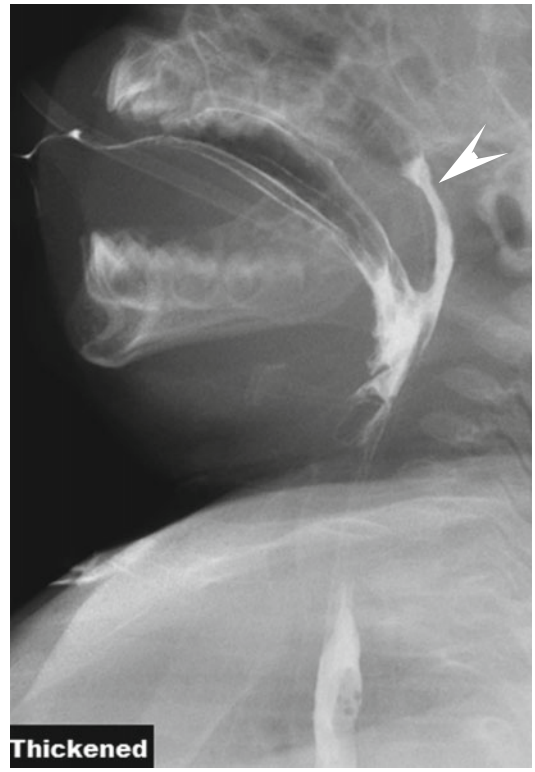

Fig. 13 Nasal regurgitation. Lateral videofluoroscopic swallow study showing nasal regurgitation (*arrowhead*)

premature infants and children with neurological disability and more commonly persist in the latter group. The causes of pediatric swallowing dysfunction (dysphagia) are varied, often complex and multifactorial in etiology, and can be either physiologic or behavioral. The most common causes of pediatric dysphagia are neurological and include cerebral palsy, traumatic brain injury, cranial nerve palsies, and meningomyelocele. Other common causes include prematurity, craniofacial abnormalities, tracheotomy, long-term ventilator dependence, and failure to thrive (Prasse and Kikano 2009). Dysphagia with aspiration is common in full-term infants less than 1 month of age and improves with age (Vazquez and Buonomo 1999). Abnormalities can occur at all anatomic levels during the oral, pharyngeal, and esophageal phases of swallowing. Micrognathia or macroglossia predisposes to oral phase dysfunction and swallowing difficulties. Children on long-term tube feeding or intensive care in the neonatal period 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 or teat. 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. 13). Occasional nasopharyngeal

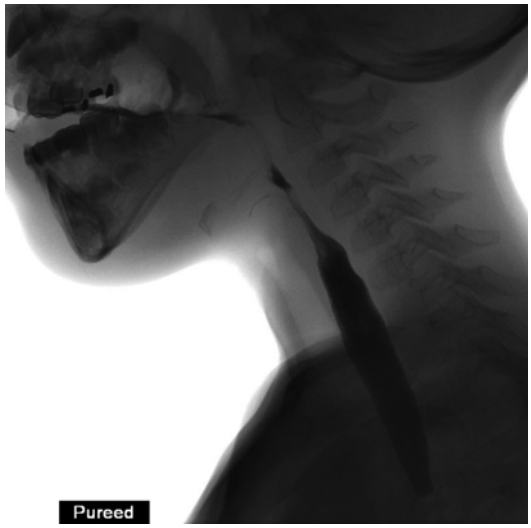

Fig. 14 Laryngeal penetration. Lateral videofluoroscopic swallow study showing trace laryngeal penetration

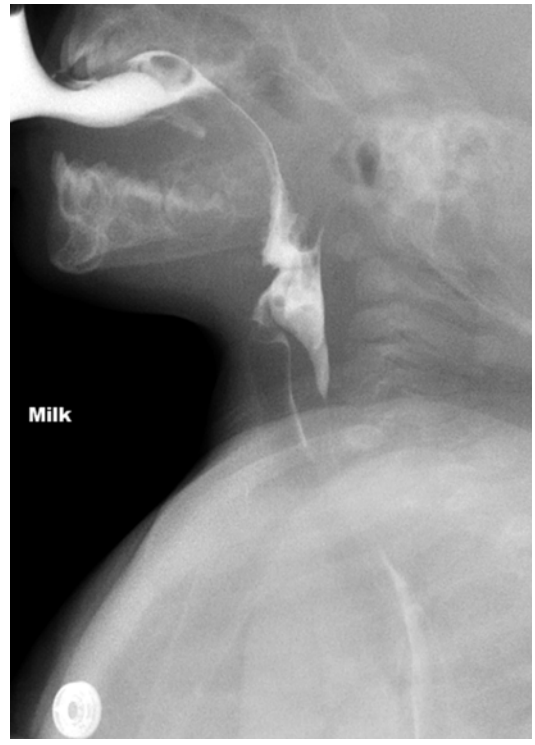

Fig. 15 Pulmonary aspiration. Lateral videofluoroscopic swallow study showing trace pulmonary aspiration

incoordination is most commonly due to transient swallowing incoordination. Retropharyngeal masses such as teratoma, lymphoma, or abscess may rarely cause dysphagia. Cricopharyngeal achalasia, a failure of relaxation of the cricopharyngeus muscle, is most commonly secondary to GER. Signs of cricopharyngeal achalasia include absent, delayed, or incomplete opening or early closure of the upper esophageal sphincter. Laryngeal penetration occurs when a bolus enters the glottis to the level of the laryngeal vestibule above the true vocal folds (Fig. 14). This may be very subtle, intermittent, or transient and referred to as "flash penetration." Pulmonary aspiration occurs when a bolus enters an incompletely protected airway beyond the level of the vocal cords and soils the trachea (Fig. 15).

3.3 Imaging Assessment of Swallowing and Esophageal Transit

The assessment of feeding and swallowing disorders in infants and children requires a multidisciplinary approach of healthcare professionals in partnership with the child's caregivers. The standard technique is the videofluoroscopic swallow study (VFSS) also referred to as the "modified barium swallow" and can help determine the consistencies of feed that a child is able to safely ingest and thereby avoid those which would predispose to aspiration. VFSS is more sensitive than combined clinical assessment of aspiration (DeMatteo et al. 2005) and conventional UGI for the detection of aspiration (Vazquez and Buonomo 1999). VFSS principally assesses oropharyngeal function and may detect related anatomical pathology. VFSS may be combined with a subsequent UGI series to assess esophageal motility and transit. A scout chest radiograph can be obtained to assess for evidence of pulmonary aspiration if there is no recent chest radiograph. Assessment of swallowing is routinely performed in young children in the true lateral position only. In older children the AP plane should also be assessed. US can be used to dynamically assess the tongue, oral cavity, hyoid, and larynx (Miller and Kang 2007) but is not widely offered as a clinical service.

VFSS requires the patient to be stationary and often the child to be securely seated. Infants can be restrained in their own child safety seat if no dedicated infant feeding seat is available. Collimation is to the hard palate, to the anterior cervical spine, and to the level of the tips of the shoulders. Orbital exposure should be avoided where possible. Unfortunately VFSS does routinely expose the thyroid to radiation. The execution of swallowing occurs too rapidly to be observed fluoroscopically. Pulsed fluoroscopy cannot be used as during routine video esophagrams as it may prevent detection of fleeting episodes of laryngeal penetration and microaspiration (Mercado-Deane et al. 2001). A video recording of the entire cinefluoroscopic study should be obtained. This can be reviewed without additional radiation exposure and compared to other historical series by the multidisciplinary team at their convenience. It is the author's practice to only save cine-loops on PACS of any abnormal series for the radiologist to review prior to any subsequent VFSS. Children may tire as feeding progresses and may have difficulty maintaining their airway so the study should be continued after the first few uneventful swallows for any given consistency (Newman et al. 2001). The child's own speech therapist or feeding therapist should ideally be present during the examination to witness the study and determine the optimal volume and order of boluses of different consistencies that are to be fed. Participation of the parents and caregivers is actively encouraged, helping to recreate some aspect of daily feeding and to reassure the child. The therapist can advise on compensatory maneuvers that assist swallowing (Fernbach 1994). Young children are given various barium-impregnated liquids of differing density depending on the type of fluid and amount of added thickener. Older children are initially challenged with a mixture of barium thickened with pudding or pureed food, yogurt, then with thin liquid barium, and finally with more solid food such as barium-coated crackers or cookies. Barium density influences transit time and 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 examination must be terminated if aspiration occurs and any changes in vital signs should be monitored with appropriate airway suctioning and supplementary oxygen administered as required. It is important to document whether aspiration induces a cough reflex. Frank aspiration should be further assessed with a post-examination chest radiograph. Coordination of swallowing improves with age. Follow-up VFSS is an effective means to monitor progress.

4 Congenital Anomalies

4.1 Esophageal Atresia and Tracheoesophageal Fistula

Esophageal atresia (EA) is congenital disorder comprising of discontinuity of the esophageal lumen. In approximately 90 % of cases, EA is associated with a communication with the trachea or bronchus which is referred to as tracheoesophageal fistula (TEF). In around 7 % of cases, there is no fistula and in the remainder no EA (Spitz 2007). EA with or without TEF is the most common congenital malformation of the esophagus with a prevalence of approximately 2.5–3.5 per 10,000 births (Pedersen et al. 2012; Shaw-Smith 2006). The underlying etiology of EA/TEF is poorly understood but is fundamentally related to disruption of separation of the embryonic proximal foregut into ventral respiratory and dorsal gastrointestinal tubes (Felix et al. 2009).

The original anatomical classification of EA was proposed by Vogt (1929) and stratified based on the presence or absence of a TEF; this was subsequently adapted by Ladd (1944) and Gross (1953). The main classification systems for EA/TEF are summarized in Table 2. Regardless, it is important to accurately describe the underlying anatomical substrate (Fig. 16) to aid preoperative planning.

4.1.1 Antenatal Diagnosis

Antenatal diagnosis of EA should be considered when there is a small or absent fetal stomach in the setting of maternal polyhydramnios and

detection of amniotic fluid-filled distended atretic esophageal pouch. The latter sign is referred to as the upper neck "pouch sign" (Kalache et al. 1998) and is only visualized in a third of cases (Fig. 17). The detection rate of EA using antenatal US is quoted at 10–40 % with a PPV of only 50 % (Hochart et al. 2015). The imaging characteristics on fetal MRI (Fig. 18) associated with the highest positive predictive values for EA are an esophageal pouch (100 %) and a small stomach (75 %) (Ethun et al. 2014).

Table 2 Summary of the different types of EA and TEF

Classification				
Vogt – Ladd type		Gross – type	Description	Frequency (%)
1			Esophageal agenesis	Rare
2	I	A	"Long-gap" or "pure" EA	7
3a	II	B	EA with proximal TEF	1
3b	III, IV	C	EA with distal TEF	86
3c	V	D	EA with both proximal and distal TEF	2
4		E	H-type TEF without EA	4
		F	Congenital esophageal stenosis	rare
			EA with distal fistula connecting to the right mainstem bronchus	rare

Adapted from Vogt (1929), Ladd (1944), Gross (1953), and Spitz (2007)

4.1.2 Postnatal Diagnosis

Salivary secretions rapidly accumulate in infants with EA who typically present with drooling, cough, or respiratory distress on attempted feeding. Children with isolated TEF (H-type type fistula) are more clinically challenging to diagnose. They may present later in life with recurrent feed-related coughing, choking, cyanotic episodes, reflux, or silent aspiration pneumonia.

4.1.3 Radiological Appearances

The radiological appearance depends on the type of lesion and whether there is esophageal atresia, patent fistula(e), or both. In most cases a chest x-ray with an orogastric tube in situ will be sufficient to make the diagnosis of EA. The diagnosis of EA should be suspected when an orogastric tube is gently advanced in a neonate and resistance is unexpectedly encountered at a relatively short placement distance. The pouch length may be inferred from a radiograph if the esophageal tube coils (Fig. 19) within the atretic pouch. However, it should be noted that the pouch is distensible and this would likely underestimate the pouch size. Care should be taken not forcibly

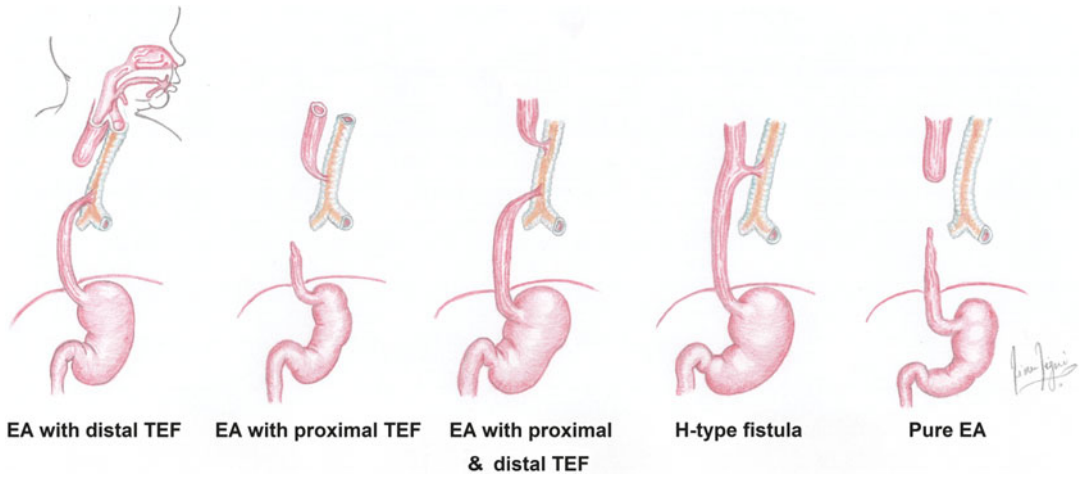

Fig. 16 Line diagram summarizing the main types of esophageal atresia (*EA*) and tracheoesophageal fistulae (*TEF*). Image courtesy of Dr. Aisha Rizvi MBBS, Doha, Qatar

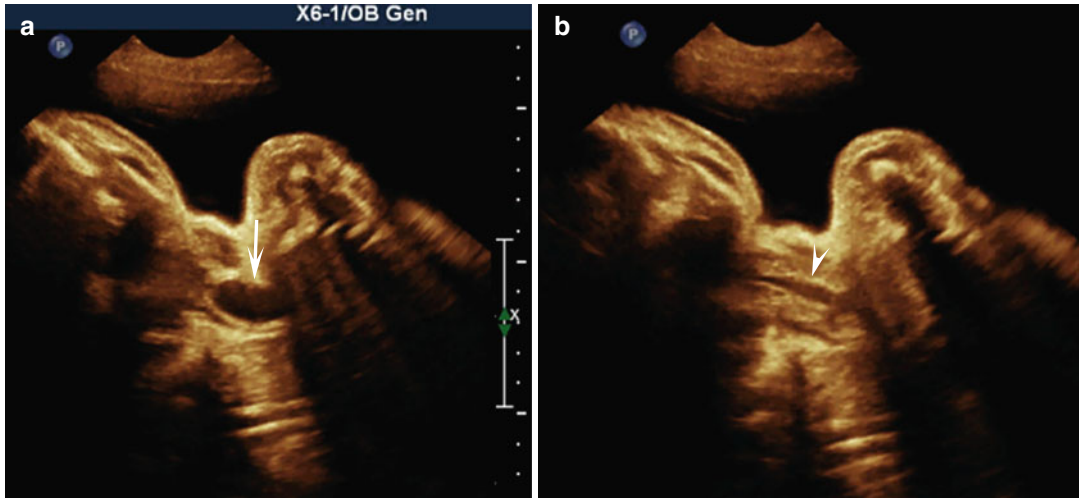

Fig. 17 (**a**, **b**) Antenatal US of esophageal atresia (EA). (**a**) Longitudinal fetal sonogram demonstrating a characteristic distended upper-pouch sign (*arrow*) in pure EA and (**b**) normal trachea (*arrowhead*). Images courtesy of Dr. Karim D. Kalache MD, Division Chief of Fetal and Maternal Medicine, Sidra Medical and Research Center, Doha, Qatar

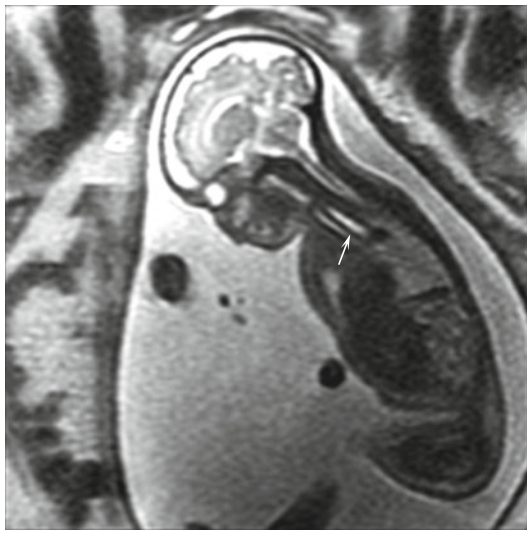

Fig. 18 Fetal MRI of esophageal atresia. Sagittal T2W HASTE image showing a fluid-filled dilated esophagus (*arrow*), polyhydramnios, and absent stomach. Image courtesy of Dr. Ashley J. Robinson MBChB, Division Chief of Interventional Radiology, Doha, Qatar

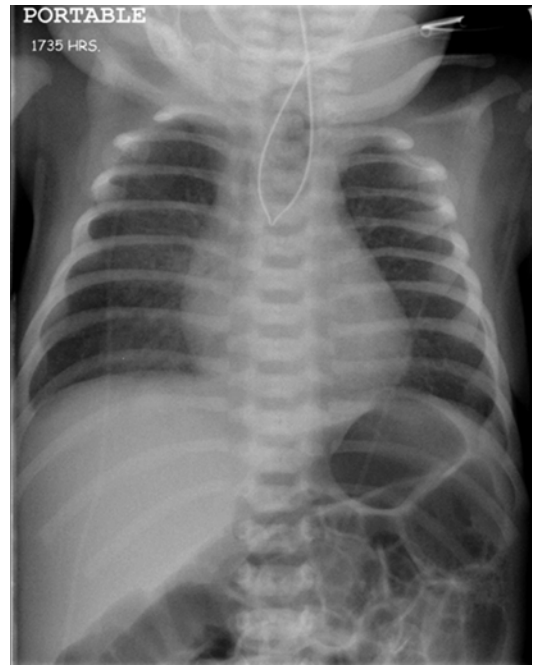

Fig. 19 Esophageal atresia with distal tracheoesophageal fistula. The orogastric tube has coiled within the atretic esophageal pouch

advance the enteric tube which may result in perforation of the atretic segment. An oral Replogle tube should be placed to decompress the esophagus and freely drain the salivary secretions to minimize the risk of aspiration. The characteristic radiological features of EA include a radiolucent, air-filled, distended, proximal blind-ending pouch (Fig. 20a). The lateral chest radiograph

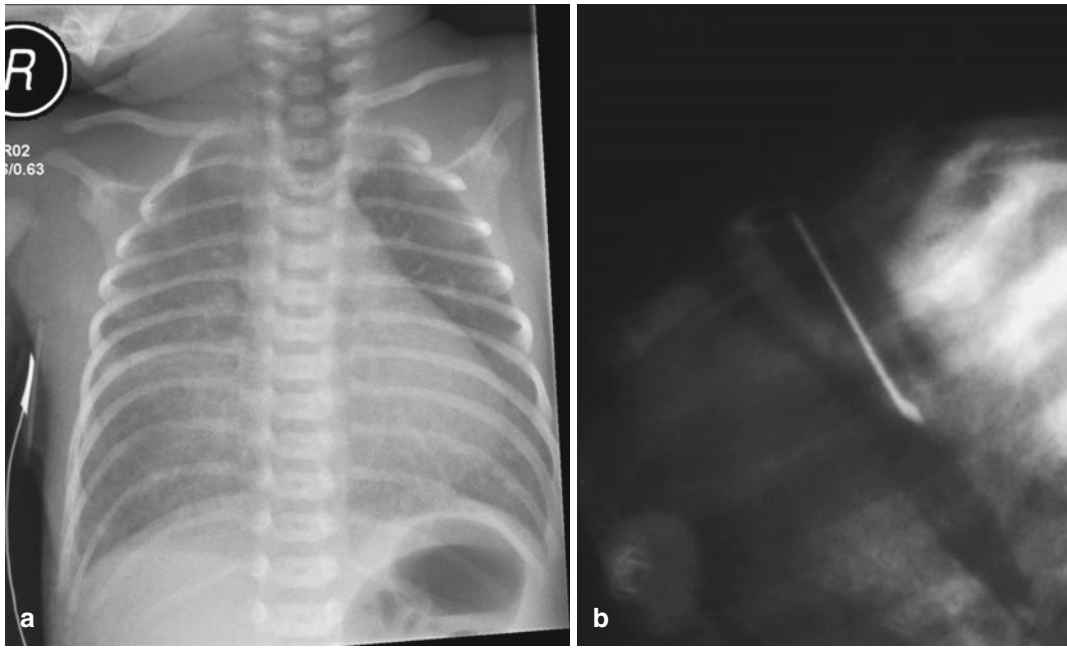

Fig. 20 (**a**, **b**) Esophageal atresia with distal. (**a**) Initial radiograph with no enteric tube shows distension of the atretic esophageal pouch. (**b**) Tracheomalacia. Lateral radiograph with the orogastric tube in the distended atretic proximal esophagus and markedly decreased caliber of the airway at the same level

confirms the distended esophageal pouch, which displaces the airway anteriorly, effacing the tracheal lumen (Fig. 20b). 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). Mechanical ventilation predisposes to gastric overinflation but can be avoided where possible by placing an endotracheal tube just beyond the most distal fistula (Spitz 2007). An initial radiograph must include the entire abdomen to assess for the presence of bowel air. A normal bowel air pattern is seen with a TEF (Figs. 21). A gasless abdomen implies the absence of a patent fistula and isolated EA (Fig. 22). The presence of 13 ribs is a predictor of long-gap atresia in TEF (Kulkarni et al. 1997).

A lateral "pouch-o-gram" of the atretic proximal esophagus is not usually necessary because of the low incidence of a fistula from the proximal pouch. A contrast pouch-o-gram can result in aspiration or respiratory compromise resulting from overdistension of the pouch and compression of the airway. Only 1–2 ml of non-ionic contrast medium or atmospheric air is required to adequately distend the pouch. Contrast medium must be removed at the end of the procedure. If primary surgical repair is deferred in a child with suspected long-gap esophageal atresia, then the patient is palliated with a percutaneous endoscopic gastrostomy (PEG). A lateral pouch-o-gram may be combined with a contrast series administered via the PEG tube to determine the esophageal gap to aid planning of definitive repair (Fig. 23). Occasionally contrast administered via a PEG will delineate a distal TEF (Fig. 24).

An H-type fistula is often very challenging to demonstrate on a contrast esophagram (Fig. 25). Where there is ongoing suspicion of TEF with a normal barium esophagram, a further contrast examination may rarely be requested, referred to as a tube esophagram. This study preoperatively has a high false-negative rate and bronchoscopy is preferable. During the tube esophagram, the infant can be placed on a board in the true lateral or prone position. The lateral position affords unobstructed views of the airway and esophagus

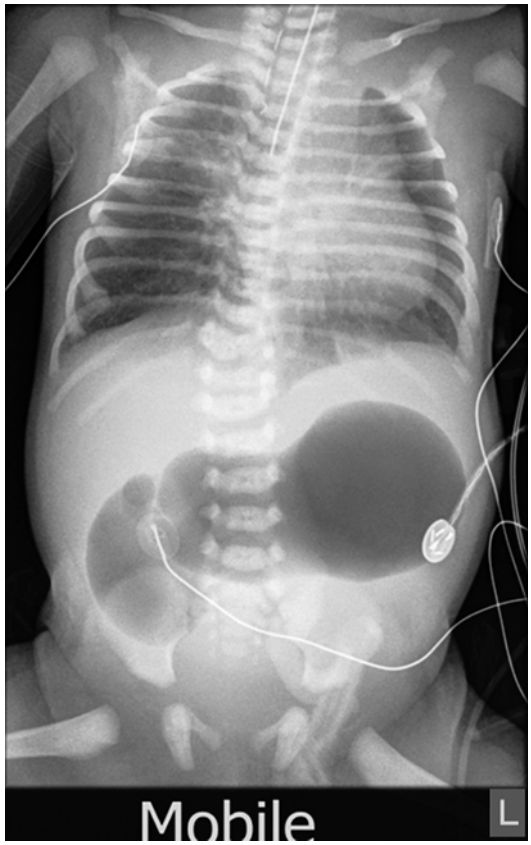

Fig. 21 Esophageal atresia with distal tracheoesophageal and VACTERL association. An oral Replogel tube is in situ. The endotracheal tube is advanced to a relatively low termination beyond the distal fistula. Lower thoracic vertebral segmentation anomalies are present. There is a characteristic *double-bubble* sign with gaseous distension of the stomach and proximal stomach due to duodenal atresia

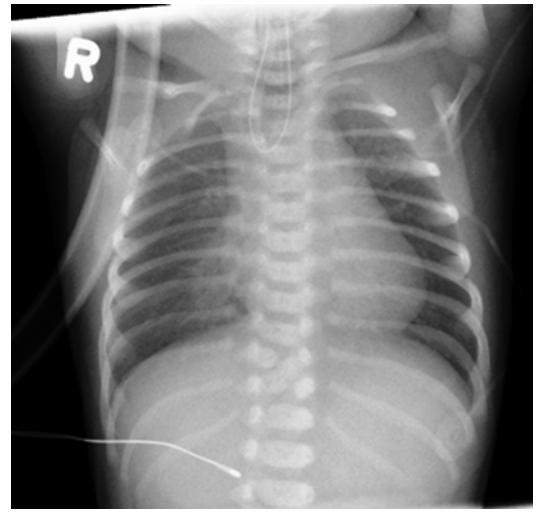

Fig. 22 Esophageal atresia without tracheoesophageal fistula. The imaged abdomen is gasless, a full length abdominal radiograph was confirmatory

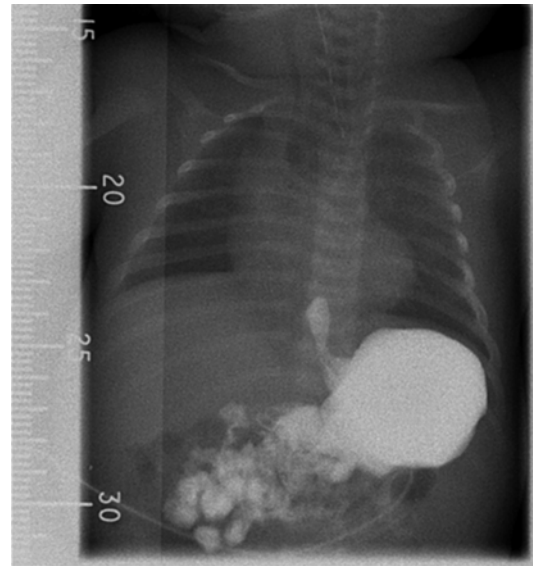

Fig. 23 Long-gap esophageal atresia. The atretic proximal esophageal pouch is distended with a small amount of air while simultaneously the stomach is opacified with contrast via a gastrostomy. The gap is just under five vertebral bodies in length

and gives the best access for suctioning. Biplane or high-resolution lateral fluoroscopic screening using a high frame rate and video capture facilities are highly desirable in order to depict a subtle fistula. The examination is performed with an enteric tube inserted into the esophagus at the level of the carina. This is slowly withdrawn while low-osmolar non-ionic contrast medium is injected at a rate sufficient to adequately distend the esophagus. The field of view should include the larynx in order to confidently differentiate between contrast entering the trachea through a subtle fistulous connection and inadvertent soiling of the airway by overflow resulting in aspiration. Great care must be exercised as the child may develop cyanosis, respiratory distress, or even apnea following inadvertent pulmonary aspiration. The upper cervical esophagus is the most common location for an H-type fistula. TEF

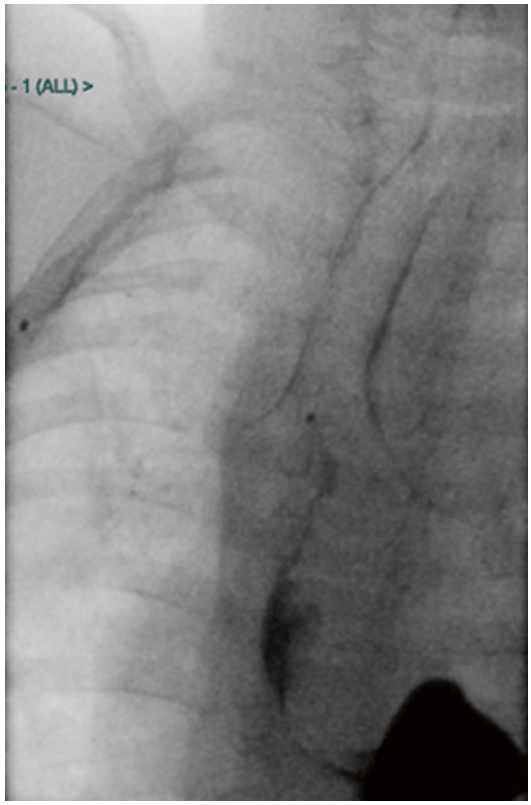

Fig. 24 Distal tracheoesophageal fistula demonstrated by contrast administered via a gastrostomy catheter

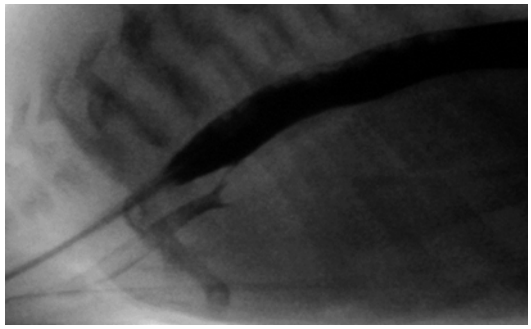

Fig. 25 Prone tube esophagram showing an H-type tracheoesophageal fistula with oblique and cephalad course

are muscular tubes that are not consistently open. TEF are thought to open with swallowing or during respiration. A fistula may temporarily occlude with food or secretions. It is not uncommon for the fistula not to be demonstrated the first time on an initial screening run on a tube esophagram.

The differential for EA includes esophageal stricture and web. The differential for TEF includes laryngotracheoesophageal cleft, esophageal diverticulum, and tubular duplications.

4.1.4 Associated Abnormalities

EA may be isolated, associated with birth defects or form part of a syndrome. There is a high frequency of congenital abnormalities associated with EA/TEF which can have significant short- and long-term implications for the child. VACTERL association is a combination of congenital anomalies typically characterized by the presence of at least three of the following components: vertebral defects, duodenal and/or anal atresia, cardiac defects, TEF, renal anomalies, and limb abnormalities. Careful physical examination should be supplemented by preoperative echocardiography in these babies to exclude structural heart disease and confirm the laterality of the aortic arch. Thoracotomy or endoscopic access is obtained on the side contralateral to the aortic arch. Radiographs should be evaluated for aortic arch laterality and vertebral and limb anomalies. US screening of the renal tract is preferably performed prior to surgery. US evaluation of the spine is also performed to exclude tethered cord but should not delay surgical repair.

Autosomal recessive disorders associated with EA/TEF include Fanconi anemia, rarely Fryns syndrome and the X-linked recessive disorder Opitz G/BBB syndrome (Stoll et al. 2009). Autosomal dominant syndromic associations of EA/TEF include CHARGE syndrome (coloboma, heart, atresia choanal, retarded growth, genital hypoplasia, ear deformities) and AEG syndrome (anophthalmia-esophageal-genital). Feingold syndrome is associated with EA (Van Bokhoven et al. 2005). Chromosomal abnormalities associated with EA include trisomy 18, trisomy 21, trisomy 13, and trisomy X mosaicism (Felix et al. 2007).

4.1.5 Postoperative Appearances of EA/TEF Repair

Surgical repair typically involves a lateral thoracotomy, TEF ligation, and primary anastomotic repair of the proximal and distal esophagus;

however, rarely minimally invasive surgery may be offered (Holland and Fitzgerald 2010). Some surgeons may advocate use of a trans-anastomotic tube. Where the baby is not conditioned for primary repair or the gap is too long, temporary palliation with cervical esophagostomy with oversewing of the distal esophagus may be indicated (Spitz 2007), a feeding PEG tube is placed for a staged repair. Following repair the child is typically ventilated and recovered in the intensive care unit with the neck flexed to reduce anastomotic tension (Holland and Fitzgerald 2010). For long-gap EA, defined as a distance of more than four vertebral bodies while under tension, the surgical options include delayed primary repair or a gastric, colonic, or small bowel esophageal replacement conduit.

Early postoperative period complications following EA/TEF repair include anastomotic leak and sepsis secondary to mediastinitis. An UGI series using non-ionic water-soluble contrast medium is routinely indicated four to seven days following primary repair to look for evidence of a leak prior to commencing feeds. The surgical anastomosis is always narrower than the previously obstructed proximal pouch and should not be confused with a stricture (Fig. 26a, b). A normal anastomosis will not impede or obstruct the flow of contrast medium. Small, contained anastomotic leaks may be managed conservatively and most will resolve but a few children will go on to develop a recurrent fistula. Up to 50 % of children with a leak will go on to develop an anastomotic stricture (Kovesi and Rubin 2004).

Almost invariably, some degree of esophageal dysmotility is encountered following EA/TEF repair. Other common long-term complications following EA/TEF repair include feeding difficulties, tracheomalacia, and GER. Esophageal stricture is by far the most frequent long-term complication following EA repair requiring interventional therapy. Treatment involves serial intermittent pneumatic dilatation (Fig. 26c) to prevent foreign body impaction from solid feeds (see Sect. 4.5). Treatment is less successful when associated with reflux (Kovesi and Rubin 2004). A (barium) esophagram should be performed with new onset of dysphagia or altered eating pattern that may indicate a stricture. Drooling and refusal to eat are often symptoms of foreign body or food impaction.

4.2 Congenital Esophageal Stenosis

This rare anomaly of unknown etiology is found in 1 in 25,000 to 1 in 50,000 live births. Congenital esophageal stenosis (CES) is characterized by a focal 2–3 mm circumferential narrowing of the esophageal lumen present from birth typically located at the junction of the middle third and distal third of the esophagus. CES may occur at any level. Very rarely CES may be multiple. Three distinct histological subtypes have been described: fibromuscular hypertrophy with dysplasia of the muscularis or submucosa, ectopic tracheobronchial cartilaginous remnants, and finally incomplete membranous diaphragm (Nihoul-Fekete et al. 1987).

CES in the neonatal period is frequently associated with EA and is contended by some authors to be a mild variant of EA (Berrocal et al. 1999). Care should be taken to evaluate the distal esophagus for a coexisting congenital stenosis. TEF is seen in a third of cases and other congenital anomalies in 17–33 % (Vasudevan et al. 2002). The diagnosis should always be considered in an infant with dysphagia or with an impacted foreign body following EA repair. The incidence of postoperative anastomotic leaks in children with TEF is higher when associated with CES (Newman and Bender 1997).

Children typically present outside the neonatal period with recurrent vomiting or food impaction on introduction of solids (Michaud et al. 2013). This may be preceded by failure to thrive and aspiration pneumonitis. A diagnosis should be considered in any case of acute dysphagia. The differential diagnosis includes strictures due to reflux esophagitis, caustic ingestion, and sequelae of surgery.

4.2.1 Radiological Findings

On fluoroscopic studies a CES lesion appears as an aperistaltic, gradual tapered narrowing, or as

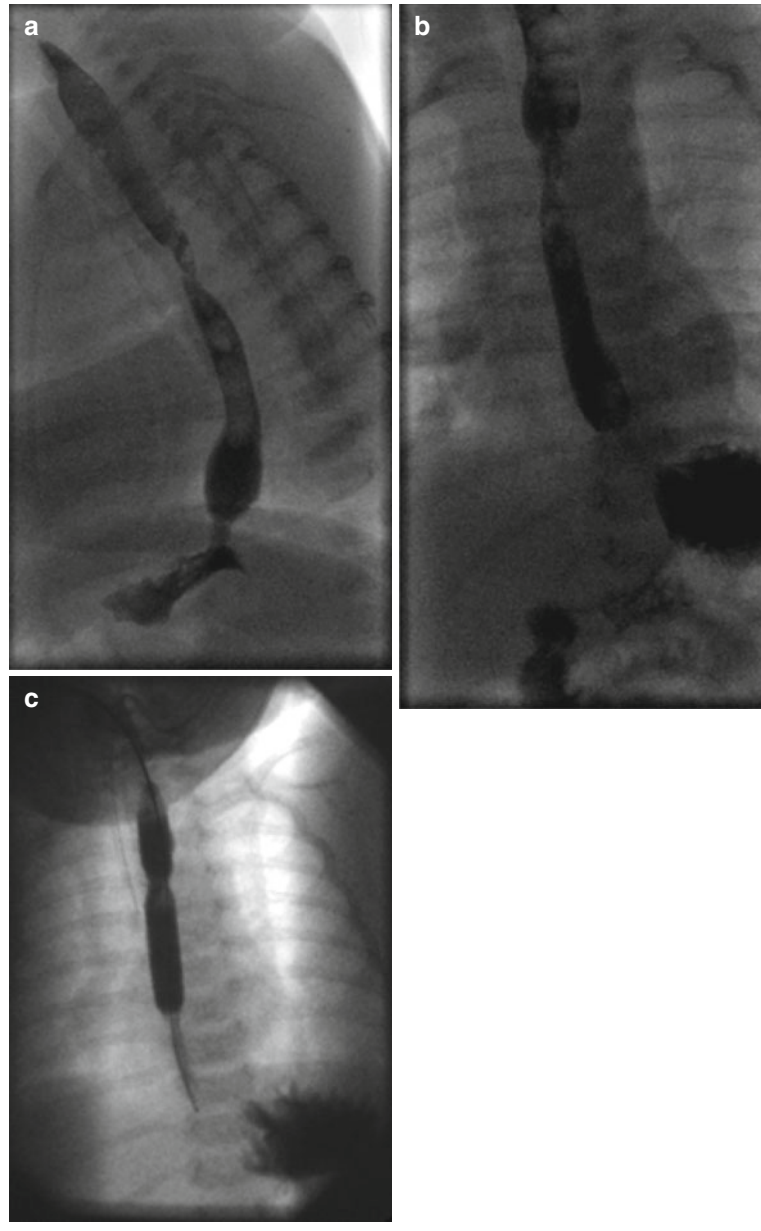

Fig. 26 (**a**–**c**) Post-anastomotic narrowing of the esophagus. (**a**, **b**) Routine postoperative swallow with non-ionic isotonic contrast medium showing esophageal narrowing at the primary anastomosis without evidence of hold up. No leak was demonstrated. (**c**) Follow-up of uneventful fluoroscopic balloon dilatation of the stricture

a persisting abrupt oblique or transverse filling defect (Fig. 27). The esophagram findings do not predict the histological subtype and endoscopic biopsy is required (Amae et al. 2003). The stenosis is smooth with intact mucosa. This may be found at the same level as a TEF. Intraluminal filling defects may represent an impacted food bolus or other foreign body. Proximal esophageal dilatation reflects a higher-grade stenosis. The entire esophagus must be carefully scrutinized after surgical correction of EA to look for coexisting CES (Vasudevan et al. 2002). Esophageal dysmotility may accompany all forms of CES.

Initial treatment may include endoscopic or interventional balloon dilatation. There is higher incidence of esophageal perforation in those with tracheobronchial remnants (TBR), especially in young children; dilatation is more effective in other histological subtypes (Michaud et al. 2013).

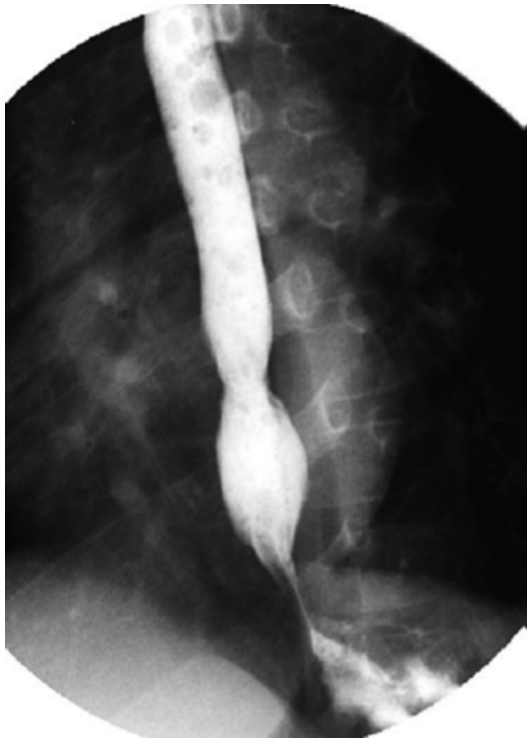

Fig. 27 Congenital esophageal stenosis. Barium swallow showing discrete annular stenosis in distal esophagus. Tracheobronchial remnants were found during histopathology

Esophageal luminal diameter increases with age and patient growth but despite repeated dilatations will never be normal (Newman and Bender 1997).

4.3 Esophageal Duplication

Esophageal duplication cysts are rare and the true prevalence is unknown. They account for approximately 15–20 % of all gastrointestinal duplications and are typically solitary. Around 80 % of cases are detected in childhood, most are symptomatic at presentation, approximately 60 % are found in the lower third of the esophagus, and the remainder evenly distributed in the proximal and middle thirds (Yoshida et al. 2005). They very rarely affect the intra-abdominal esophagus. Esophageal duplications are defined by the presence of (1) attachment to the esophageal wall, (2) gastrointestinal epithelial lining, and (3) a smooth muscle wall. Spherical duplication cysts are the product of a foregut cystic malformation and are enteric cysts which abnormally pinch off from the foregut and incorporate in the wall of the developing gastrointestinal tract (Sharma et al. 2009). They may contain gastric mucosa, which can cause mural ulceration, bleeding, and rarely perforation. Secondary infection is uncommon. The differential is a bronchopulmonary foregut cyst. Spherical esophageal duplication cysts are treated by excision. Tubular duplications represent true microscopic and macroscopic duplication with a blind loop of the gut lying along the antimesenteric border. These typically communicate with the bowel lumen. Complete esophageal duplication is extremely rare and is often associated with gastric duplication (Herman et al. 1991). Tubular duplications require more complex surgical management.

4.3.1 Radiological Findings

Esophageal duplication cysts may be detected incidentally on chest radiographs as a posterior mediastinal mass. An esophagram demonstrates a smooth, well-defined extrinsic soft tissue mass displacing the esophagus. US may adequately demonstrate a distal esophageal cyst (Fig. 28). Cross-sectional imaging enables differentiation of the cyst from a solid mass such as neuroblastoma or pulmonary sequestration and to show its relationship to adjacent vital structures. CT reveals a well-defined round or ovoid fluid containing non-enhancing mass adjacent to the esophagus (Fig. 29). On MR cyst content reflects the amount of water and proteinacious material, with signal typically following water but may appear more complex if complicated by hemorrhage.

4.4 Esophageal Bronchus

Esophageal bronchus is a rare form of congenital bronchopulmonary foregut malformation (CBPFM) involving an abnormal cartilaginous fistulous connection between the foregut and an isolated portion of respiratory tissue. The most common variant is the esophageal bronchus whereby a portion of lung communicates with

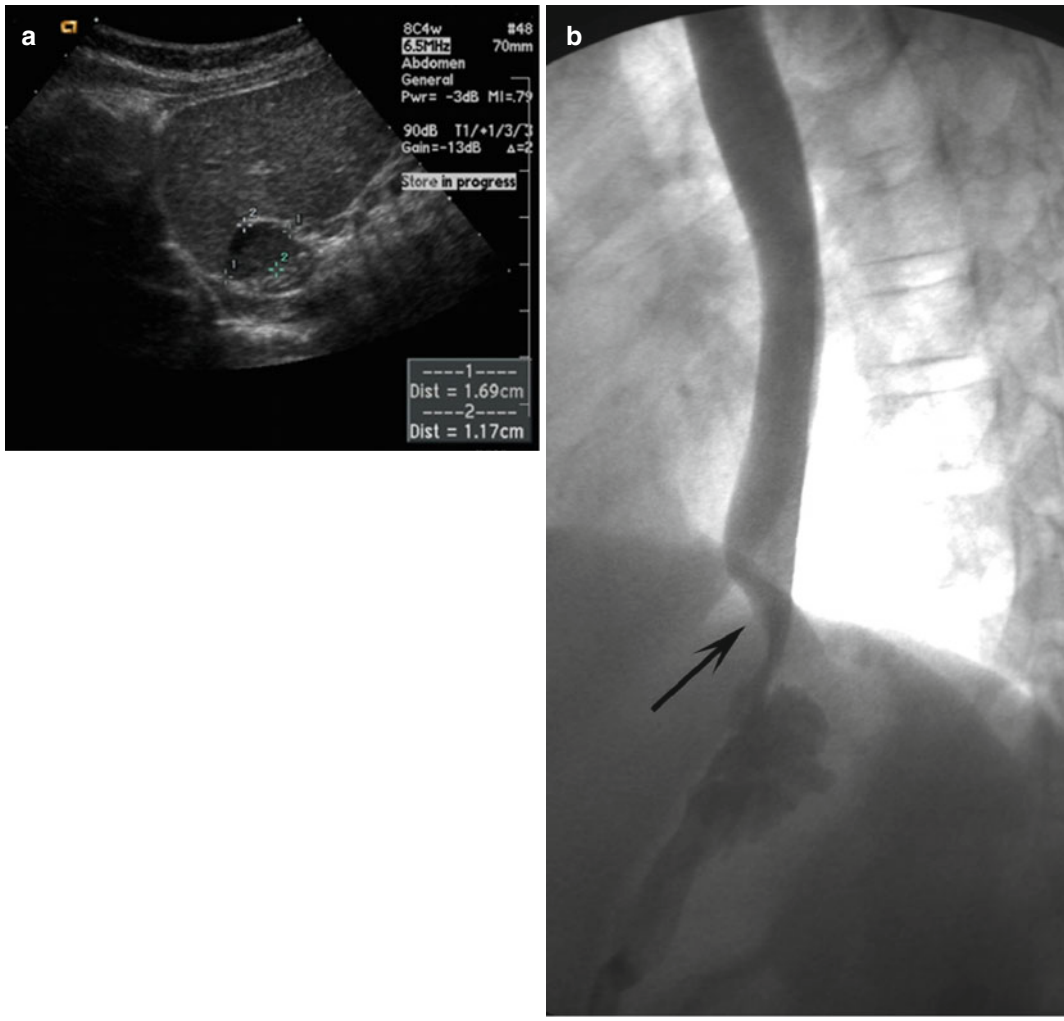

Fig. 28 (**a**, **b**) Esophageal duplication cyst. (**a**) US image showing a thin-walled subdiaphragmatic cyst (between calipers) causing (**b**) smooth extrinsic compression of the distal esophagus as shown on a barium esophagram (*black arrow*)

the lower esophagus. The anomalous communication may involve a whole lung. Involvement of the stomach is rare. The embryological classification of these fistulae is summarized in Table 3. The fistula forms through a focal mesodermal defect when a portion of the lung bud covered by mesenchyme connects with the elongating foregut (Srikanth et al. 1992). The affected bronchus is absent from the native bronchial tree. The arterial supply of the affected lung arises from the pulmonary artery or both pulmonary and systemic circulations. Venous drainage is to the pulmonary vein, azygos, or portal venous system. There is a frequent association with other bronchopulmonary foregut malformations such as congenital pulmonary airway malformation, sequestered lung, and bronchogenic cyst. Other common associations include TEF with EA and cardiac anomalies which substantially increase morbidity and mortality. Symptoms include respiratory distress, coughing related to feeds, and recurrent lower respiratory tract infections. Whole lung involvement tends to present in the neonatal period, whereas lobar involvement may present later in life. Neonatal tracheal reimplantation is the treatment of choice. However, resection of the anomalous pulmonary tissue and fistula with

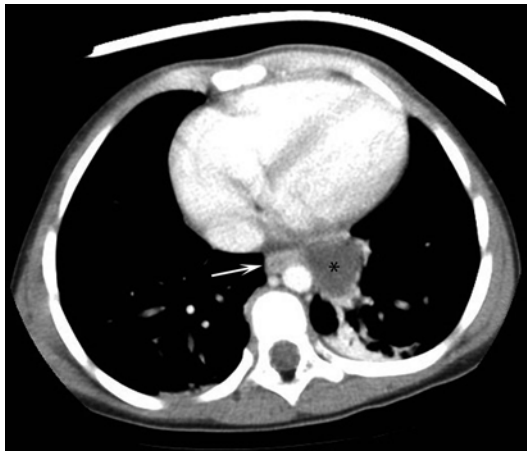

Fig. 29 Axial CT of esophageal duplication cyst (*) adjacent to the esophagus (*arrow*) and descending thoracic aorta

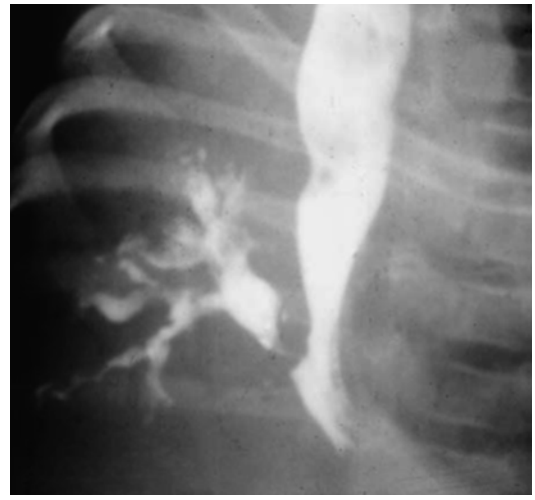

Fig. 30 Esophageal bronchus. Barium esophagram demonstrates the right main bronchus arises from the esophagus. The right lung is completely opacified

Table 3 Proposed classification of bronchopulmonary foregut fistulae

Type	Subtype	
1		CBPFM associated with esophageal atresia and tracheoesophageal fistula
	A	Entire lung arises from the esophagus or stomach
	B	Portion of one lung or lobe arises from the esophagus
2		A lung originates from the distal esophagus
3		Isolated anatomic lung, lobar or segmental fistula to esophagus or stomach
4		Portion of the normal bronchial system communicates with the esophagus and receives systemic blood supply

Adapted from Srikanth et al. (1992)

repair of the anomalous communication is required if the affected lung has been destroyed by recurrent infection (Sugandhi et al. 2011).

4.4.1 Radiological Findings

The chest radiograph is nonspecific, revealing opacification of the affected portion of lung. A water-soluble contrast examination of the esophagus and stomach using isotonic non-ionic contrast medium is sufficient to confirm an abnormal communication (Fig. 30). Unenhanced CT will reveal pulmonary parenchymal complications including atelectasis, bullae, and abscess formation and estimate percentage lung involvement. MDCT angiography (CTA) is preferable to depict adequacy of arterial blood supply and venous drainage.

4.5 Hiatal Hernia

A hiatus hernia is defined by the abnormal migration of an intra-abdominal structure in to the thoracic cavity through the esophageal hiatus. Hiatus hernia and intra-thoracic stomach are uncommon in children. They may be congenital or acquired. Most are sporadic. Associations include a short esophagus, Marfan's syndrome, and visceral heterotaxy (Al-Assiri et al. 2005). The anatomical classification of hiatal hernias is summarized in Fig. 31 and is as follows: Type I hiatal hernias, also referred to as sliding hiatal hernias, are by far the most common type. Type I hernias result from circumferential laxity of the phrenoesophageal membrane such that the gastroesophageal junction and a portion gastric cardia migrate through the widened muscular hiatal tunnel (Kahrilas et al. 2008). The

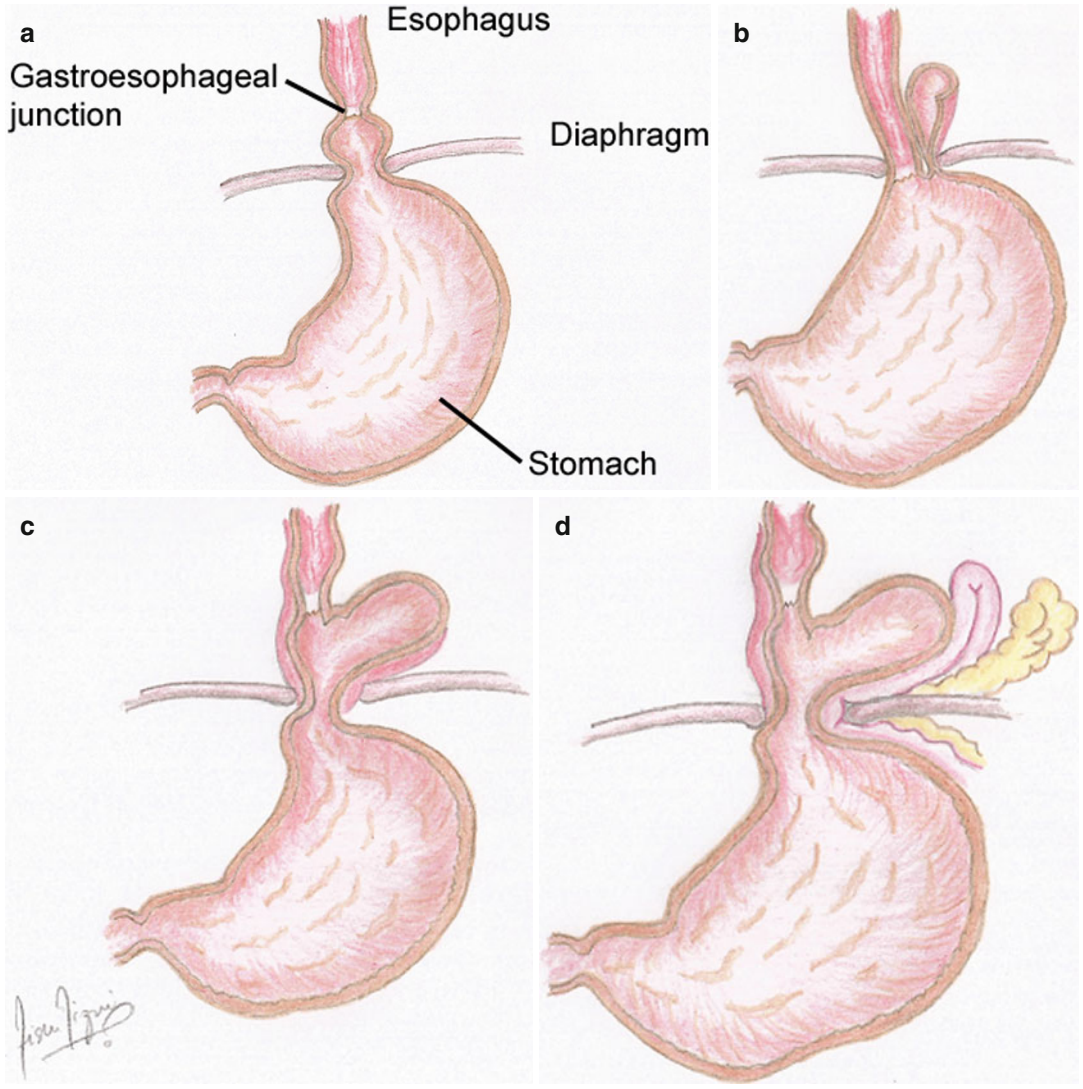

Fig. 31 (**a**–**d**) The classification of hiatal hernias. (**a**) Type I (sliding) is the most common whereby the gastroesophageal junction herniates beyond the gastroesophageal junction. (**b**) In type II hernias the gastric fundus is the lead point for herniation and the gastroesophageal junction is normally positioned. (**c**) Type III is a mixture of types I and II. (**d**) Type IV hernias involve herniation of the stomach and other organs. Image courtesy of Dr. Aisha Rizvi MBBS, Doha, Qatar

stomach remains in its usual longitudinal alignment and the fundus remains below the gastroesophageal junction. These may be intermittent and are often found incidentally on an esophagram but can predispose to GER. Type II–IV hernias are referred to as paraesophageal hernias. Type II hernias occur when part of the gastric fundus herniates through the diaphragmatic hiatus adjacent to the esophagus. The gastroesophageal junction remains in normal anatomical position. Type III hernias are the most common type of paraesophageal hernia and are a combination of Types I and II, with the fundus lying above the gastroesophageal junction. Type IV hernias are characterized by the presence of a structure other than stomach such as colon, spleen, small intestine, or pancreas in the hernial sac.

Esophageal transit time and duration of GER are prolonged in children with hiatal hernia. In symptomatic children with recurrent gastroesophageal reflux disease, laparoscopic fundoplication is advocated (Bansal and Rothenberg 2014).

4.5.1 Radiological Findings

Depending on the size of the herniated portion of the intra-thoracic stomach, chest radiographs may reveal a air-filled lucency occasionally with compressive atelectasis of adjacent lung. An esophagram is confirmatory (Fig. 32). Ultrasound can demonstrate a short length of abdominal esophagus, loss of the acute gastroesophageal angle, and GER as well as transhiatal passage of omental fat (Westra et al. 1990; Koumanidou et al. 2004).

4.6 Vascular Rings and Sling Abnormalities

4.6.1 Vascular Rings

A "vascular ring" refers to an anomaly of the embryonic aortic arch complex and involves the paired 4th–6th aortic arches. Abnormal regression or persisting patency of these arches results in vascular and ligamentous derivatives encircling and compressing the trachea and esophagus. Vascular rings are uncommon but approximately one fifth are associated with congenital cardiac anomalies. The presentation of a vascular ring is varied, but predominantly related to the degree of airway compression. Symptoms include life-threatening respiratory compromise in the infant, stridor, and recurrent chest infections. Gastrointestinal symptoms such as dysphagia, particularly on the introduction of solids, are less common and are more often encountered with a left retroesophageal subclavian artery arising from a right aortic arch (Bonnard et al. 2003). Vascular rings may often be asymptomatic and detected as an incidental finding on a contrast esophagram or cross-sectional imaging. Rarely, ingested foreign bodies may declare a vascular ring as the foreign body will lodge just proximal to the ring (see Sect. 5.3).

Approximately 75 % of cases of double aortic arch are typically formed by a dominant superior right aortic arch and completed by a more inferior smaller or atretic left aortic arch. In approximately 20 % the left aortic is dominant (Fraser and Carberry 2012). A double aortic arch results in anterior and bilateral airway compression and posterior and bilateral indentation of the esophagus. A right aortic arch with aberrant left subclavian artery (LSCA) will give rise to a vascular ring if the left ductus arteriosus passes between the right descending aorta and the left pulmonary artery and results in posterior oblique compression of the esophagus. The aberrant LSCA will often arise from a diverticulum of Kommerell.

The traditional surgical treatment of a vascular ring is to relieve the constriction by open thoracotomy with division of non-functional or non-critical components of the ring, typically the atretic or nondominant segment (Alsenaidi et al. 2006). In cases with a right aortic arch and a left ligamentum arteriosum, the ductal remnant is divided, and the trachea and esophagus are released from fibroadhesive bands (Backer et al. 2005). Primary translocation of the aberrant left subclavian artery to the left carotid artery, with removal of the Kommerell diverticulum and division of the ligamentum through a left thoracotomy, is an alternative strategy (Shinkawa et al. 2012). Minimally invasive thoracoscopic division of a vascular ring avoids the morbidity associated with thoracotomy (Al-Bassam et al. 2007).

4.6.1.1 Radiological Findings

A chest radiograph may demonstrate tracheomalacia and confirm the laterality of the aortic arch(es). A right-sided aortic arch in a child with respiratory symptoms is suspicious for a vascular ring. The next examination is typically a (barium) esophagram which best depicts extrinsic esophageal compression by an atretic or persistent aberrant vascular segment which are typically not well delineated by MDCT or MRI (Turner et al. 2005). The esophagram is a useful screening modality as it is widely available, may exclude other esophageal pathologies, and does not require sedation. An adequately distended normal esophagram will exclude a vascular ring.

A vascular ring caused by a double aortic arch system with bilateral arch patency causes significant focal narrowing and anterior indentation of

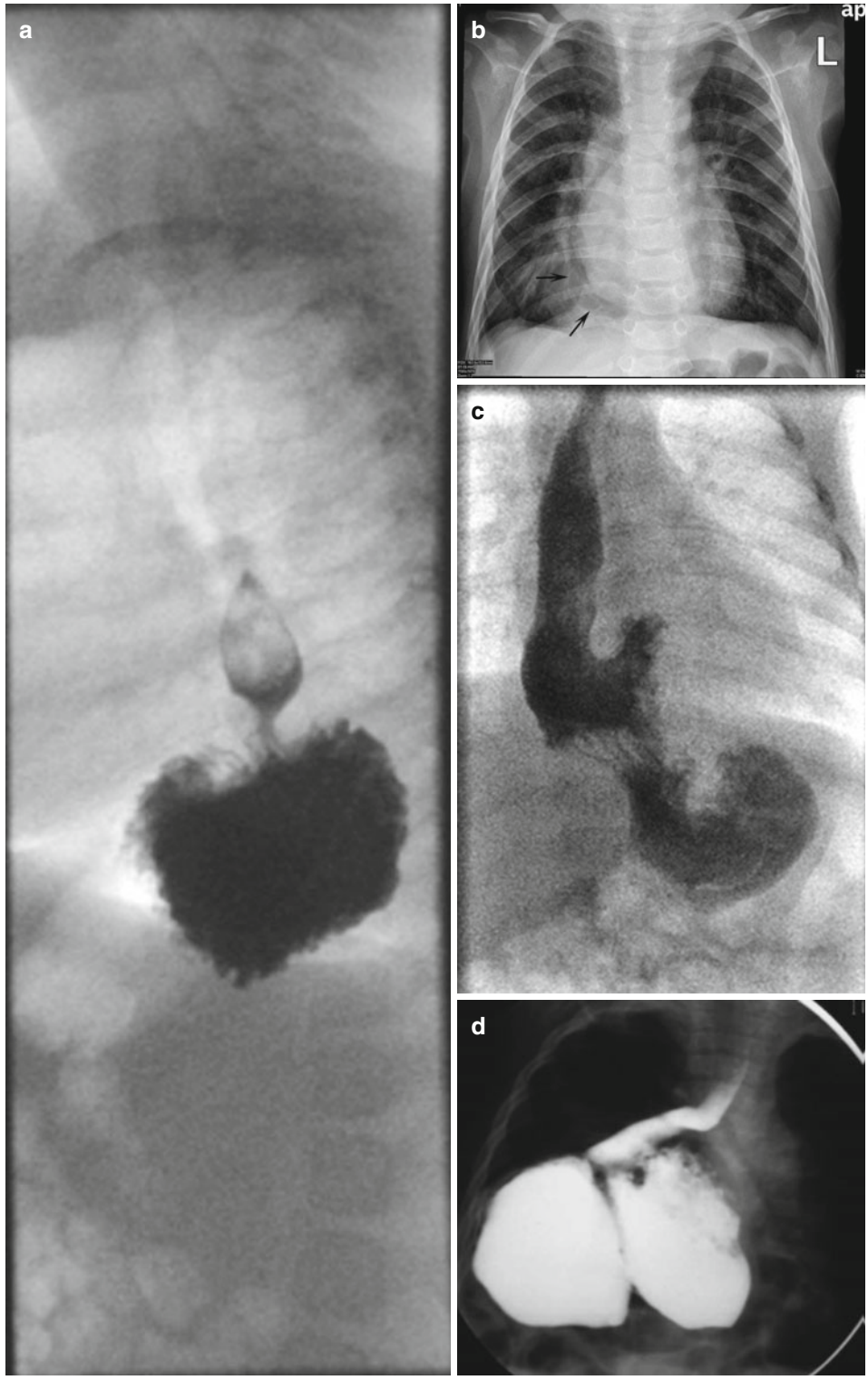

Fig. 32 (**a**–**d**) Radiographic imaging of hiatal hernias. (**a**) Barium swallow demonstrating a type I hiatus hernia. (**b**) Frontal chest radiograph showing a retrocardiac lucency (*arrows*) (**c**) confirmed on a barium swallow to be a type III hiatus hernia. (**d**) Intra-thoracic herniation of the stomach in a child with Marfan's syndrome

the trachea, which can be appreciated on the lateral chest radiograph and barium esophagram (Fig. 33a, b). A characteristic S-shaped configuration of right- and left-sided lateral indentations is present on the frontal projection. A barium esophagram can depict the right arch and a posterior esophageal impression that is frequently associated with congenital heart disease. A contrast

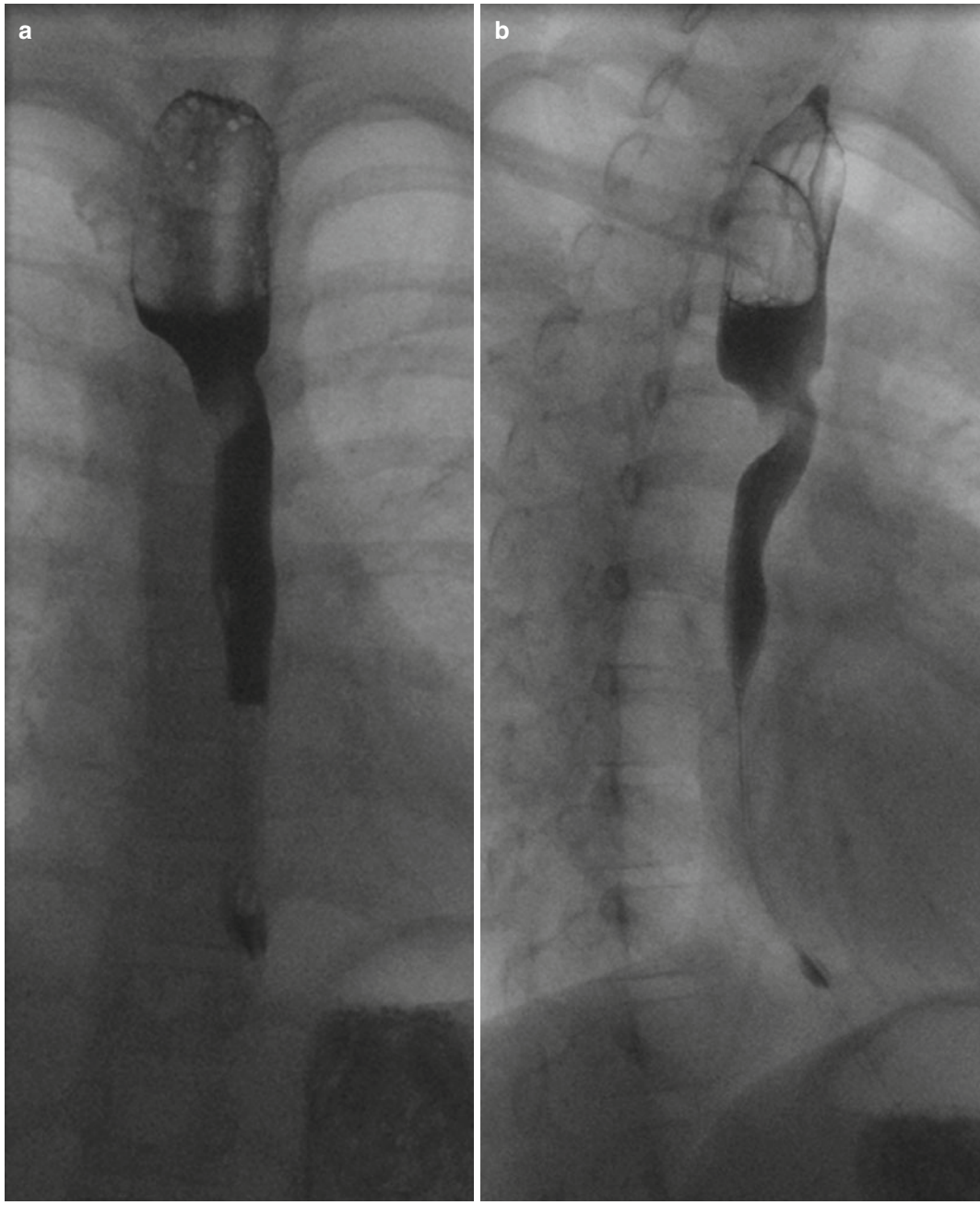

Fig. 33 (**a**–**e**) Double aortic arch. Barium esophagram (**a**) AP view showing bilateral extrinsic impressions upon the esophagus and (**b**) the lateral view showing a posterior indentation. (**c**) Axial contrast-enhanced maximum-intensity projection CT showing a complete double aortic arch encircling the trachea and esophagus, (**d**) corresponding 3D volume-rendered image and (**e**) coronal minimum-intensity projection image showing tight compression of the trachea

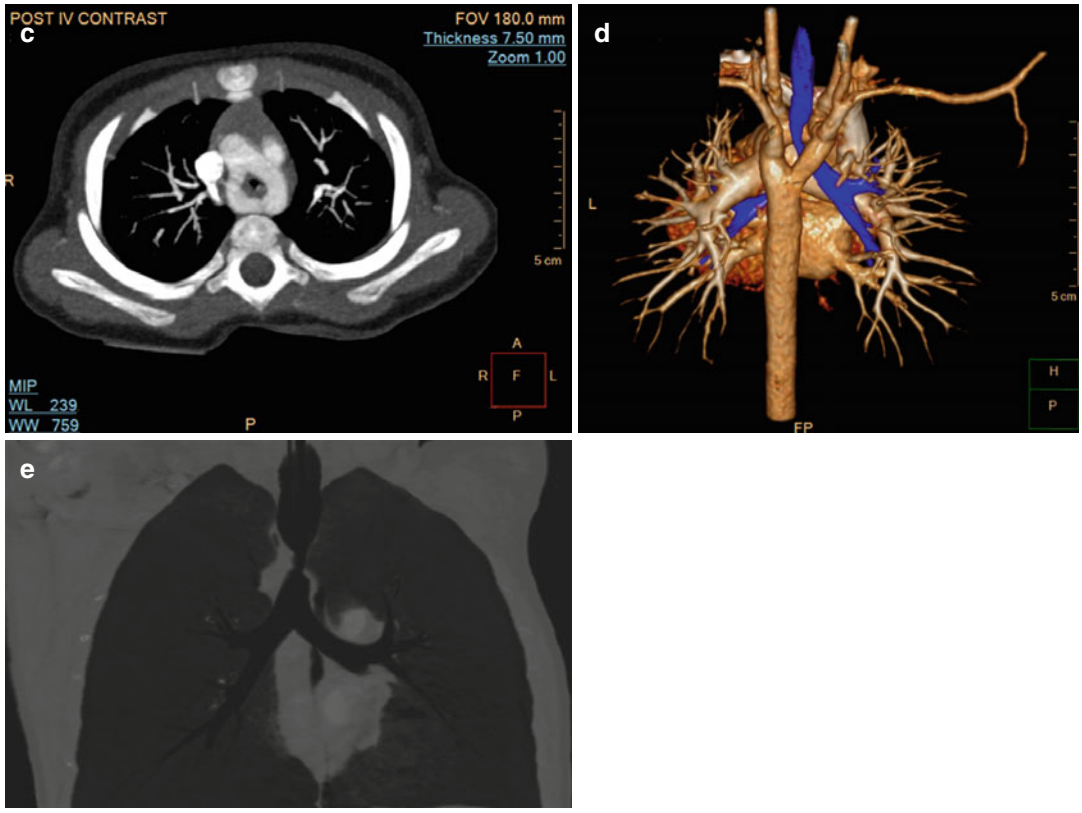

Fig. 33 (continued)

esophagram does not reliably distinguish between the various other forms of vascular ring. Cross-sectional imaging, either with MR or enhanced MDCT, is most useful delineating the spatial anatomy to provide a preoperative roadmap for thoracoscopy or an open thoracotomy (Yedururi et al. 2008). MDCT is rapidly acquired over a few seconds without the need for sedation or anesthesia in most cases (Fig. 33c–e). MR typically would require sedation and immobilization for studies that can take between 45 and 60 min. Respiratory and EKG-navigator MRI with feeding and swaddling is feasible for the very young neonate to avoid sedation. The obvious disadvantage of MDCT is the radiation dose and pediatric-specific protocols are mandated. MDCT is still favored by many as the lungs and bronchi are exquisitely demonstrated. Diagnostic yield is further enhanced with multiplanar, maximum-intensity projection, minimum-intensity projection, and 3D volume-rendered reformations. High-resolution black blood, 3D FISP (fast imaging with steady precession), and magnetic resonance imaging (MRA) will accurately define vascular rings (Fig. 34). The choice of imaging modality varies with institutional preference.

4.6.2 Pulmonary Artery Sling

Pulmonary artery sling complex is a rare vascular anomaly in which the left pulmonary artery (LPA) arises from the right pulmonary artery, encircles the distal trachea and right main bronchus, and indents the anterior margin of the esophagus prior to entering the left lung hilum. The LPA ring-sling complex is defined as the association of an aberrant LPA with complete cartilaginous tracheal rings and tracheal stenosis. Symptoms are typically related to airway compression. Surgical repair requires relief of the sling compression with reimplantation of the LPA to the main pulmonary artery, while tracheal rings require major airway reconstruction with a slide tracheoplasty.

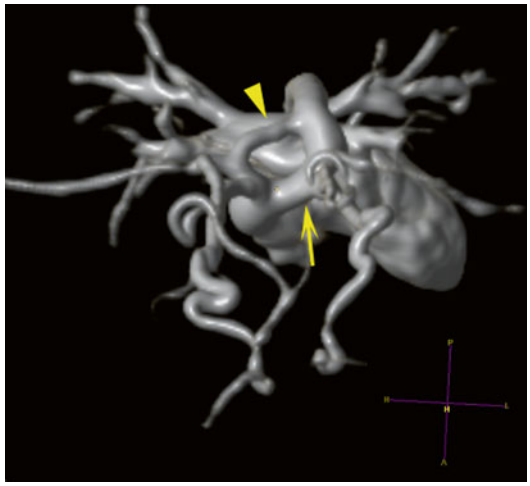

Fig. 34 Double aortic arch. Contrast-enhanced MRA of the thorax in a child demonstrating a dominant left aortic arch (*arrow*) and smaller right arch (*arrowhead*)

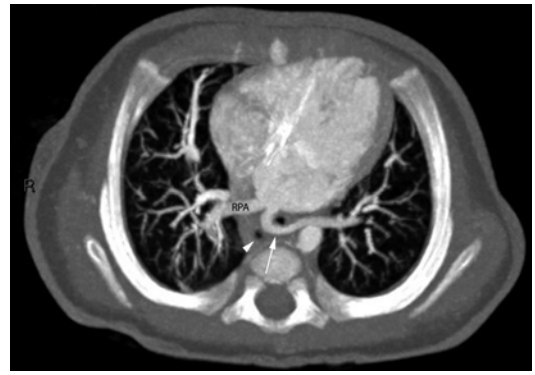

Fig. 35 Anomalous left pulmonary artery origin. Axial contrast-enhanced CT maximum-intensity projection image showing anomalous origin of the left pulmonary artery (*arrow*) from the right pulmonary artery (*RPA*). The LPA passes behind the trachea and forms an anterior indentation upon the esophagus (*arrowhead*)

4.6.2.1 Radiological Findings

Cross-sectional imaging with MDCT (Fig. 35) or MR is helpful in presurgical planning and post-surgical assessment. A barium esophagram is not usually performed but may reveal an abnormal anterior indentation upon the esophagus by the anomalous LPA.

5 Acquired Abnormalities

5.1 Gastroesophageal Reflux

Gastroesophageal reflux (GER) is defined by the retrograde passage of gastric content into the esophagus with or without vomiting or regurgitation. GER is very common in infants and children and may be physiologic in young infants. Gastroesophageal reflux disease (GERD) refers to those children with GER who develop troublesome symptoms and/or complications (Vakil et al. 2006).

The incidence of GER ranges from 25 to 60 % in all infants (Rosen 2014) and up to 70 % in children with underlying conditions such as tracheoesophageal fistula, neurological deficits, and anatomic abnormalities of the esophagus (McGuirt 2003). The clinical presentation of GER is variable. In infants it manifests as feeding difficulties including vomiting, regurgitation, and 'spitting up'. Other symptoms include arching, hoarseness, and cough. GER has a peak incidence at 4 months of age, decreasing by 6 months and falling sharply by around 12 months of age coinciding with the age at which children learn to sit up (Campanozzi et al. 2009). Epigastric pain is more common in older children. In young infants, the short length of the intra-abdominal esophagus and physiologic immaturity of the developing lower esophageal sphincter (LES) contribute to GER. A transient fall in LES pressure to a level at or below intragastric pressure results in GER. GER invariably improves with the introduction of solid food. Severe GERD can lead to growth disturbance and troublesome gastroesophageal symptoms such as "heartburn" and refusal to eat. GER is associated with asthma. Hyperinflation changes the pressure gradient across the LES, increases negative intra-thoracic pressure, and alters the relationship between the diaphragm and lower esophageal sphincter. This may be exacerbated by some asthma medications that decrease LES pressure. Pulmonary aspiration is a significant complication although seldom demonstrated by diagnostic imaging.

Clinical investigation of GER includes a 24-h continuous ambulatory esophageal pH-impedance test to document acid and non-acid reflux events. pH studies have a much higher

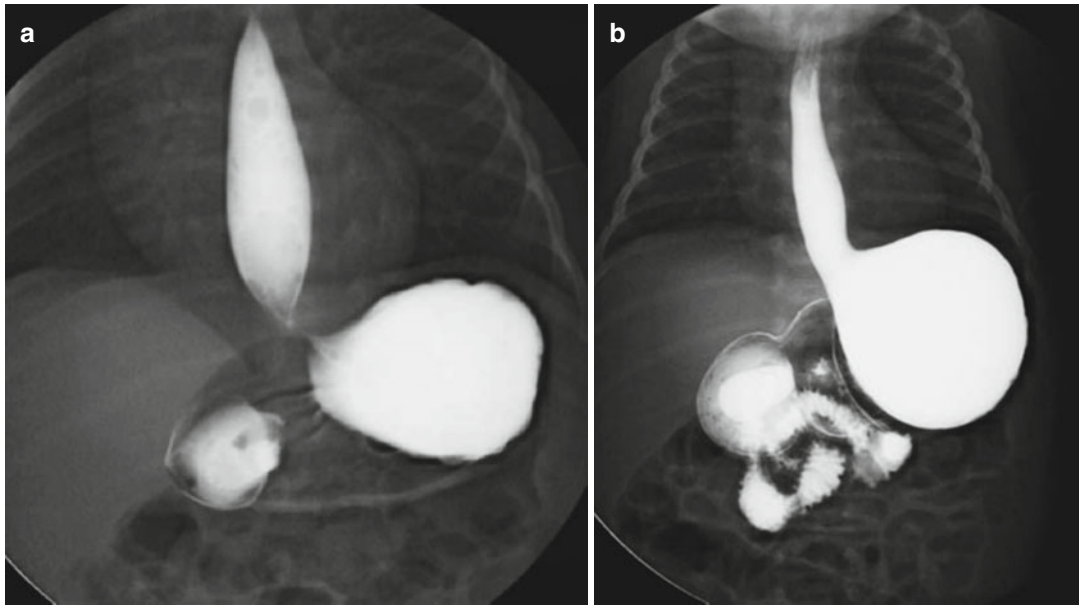

Fig. 36 (**a**, **b**) Gastroesophageal reflux. (**a**) During swallowing, the gastroesophageal junction is closed. (**b**) The gastroesophageal junction is widely patent, and barium refluxes to the upper esophagus

sensitivity for GER than radiological investigations (Vandenplas et al. 2009). pH studies without impedance are more widely available but do not document significant non-acid reflux episodes and rely on the period of time pH falls below 4, referred to as the pH index. Endoscopy will confirm macroscopic features of GERD such as esophagitis, erosions, ulceration, and stricturing with the potential for biopsy. Esophageal manometry detects LES pressure and may diagnose achalasia of the cardia or transient relaxation of the LES implicated in GERD. All these techniques are highly invasive.

The initial treatment for GERD is conservative and most commonly involves a regime of offering smaller boluses of thickened feed and maintaining an upright position after feeding. Children with GERD refractory to medication may benefit from surgery. Laparoscopic fundoplication whereby the LES is augmented with a gastric fundal wrap is the surgical treatment of choice for GERD. In some children the wrap can be "too tight" leading to troublesome dysphagia and may require endoscopy +/− dilatation (Kubiak et al. 2011). Fundoplication failure requiring reoperation is uncommon but most common in children with neurological disability.

5.1.1 Radiological Findings

The (barium) esophagram and UGI series is frequently requested in children with suspected GER (Fig. 36) and is the least invasive routine diagnostic imaging test. Contrast studies have low sensitivity (43 %) and low negative predictive value (24 %) for detecting GER (Macharia 2012). The main role of the contrast examination is to exclude anatomic abnormalities of the esophagus such as hiatal hernia, to define the position of the duodenojejunal junction; document gastric emptying, the cephalad anatomical level of any reflux, evaluate for pulmonary aspiration and assess the effectiveness of a fundoplication procedure (Fig. 37). Late complications of GERD in childhood include esophagitis, stricture formation (Fig. 38), and Barrett's intestinal metaplasia.

Dilute barium is administered orally. Water-soluble contrast medium may be initially used if there is initial concern for aspiration. Ideally the stomach should be filled with the same volume as a normal feed. If barium intake is insufficient, the ingested volume can be supplemented with infant formula or fruit juice. Chilled barium and adding flavorings may improve palatability. Alternatively

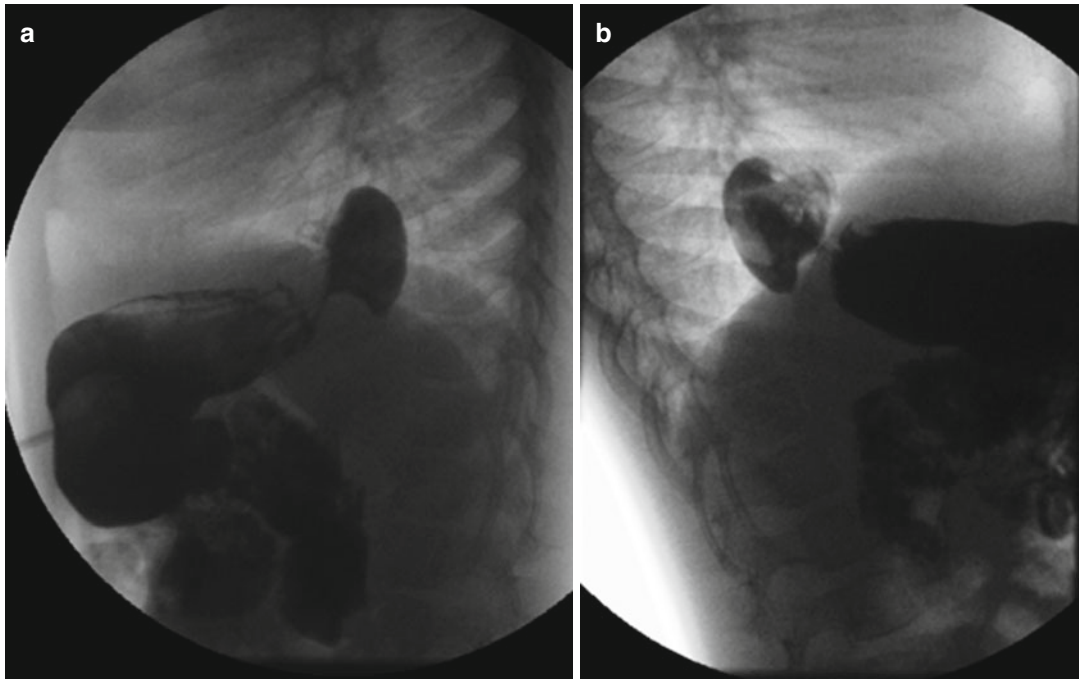

Fig. 37 (**a**, **b**) Nissen fundoplication. (**a**) Lateral and (**b**) AP view of an esophagram showing the subdiaphragmatic wrap. Adequate passage of contrast through the fundoplasty into the stomach is demonstrated

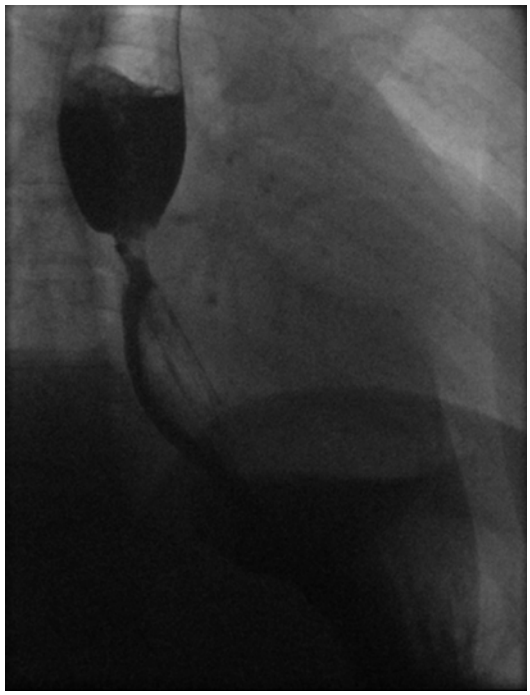

Fig. 38 Reflux stricture. Tapered stricture of the distal esophagus in a teenager with GERD refractory to medical therapy

the feed may be administered via an indwelling percutaneous gastrostomy tube. The stomach can be filled via a nasogastric tube. As any enteric tube passing through the GEJ maintains patency of the lower esophageal sphincter and impairs its function, such tubes must be immediately removed when evaluating for GER. The presence of GER does not necessarily imply GERD nor does its absence exclude GERD (Rudolph et al. 2001). Imaging for GER could be construed as "objectifying the subjective." The estimated dose of an UGI series in a single center using 16 spot images and 40 s of fluoroscopy is of the order of around 1 mSv and almost equivalent to annual background radiation exposure (Macharia 2012).

The radionuclide "milk scan" is a sensitive test for diagnosing GER but is not routinely indicated for assessing GERD (Vandenplas et al. 2009). Milk, formula, or juice mixed with Tc 99 m sulfur colloid is administered to the child who is then scanned. Scintigraphy offers continuous dynamic assessment enabling documentation of the rate of gastric emptying, number of episodes of GER, and cephalad extent of each GER

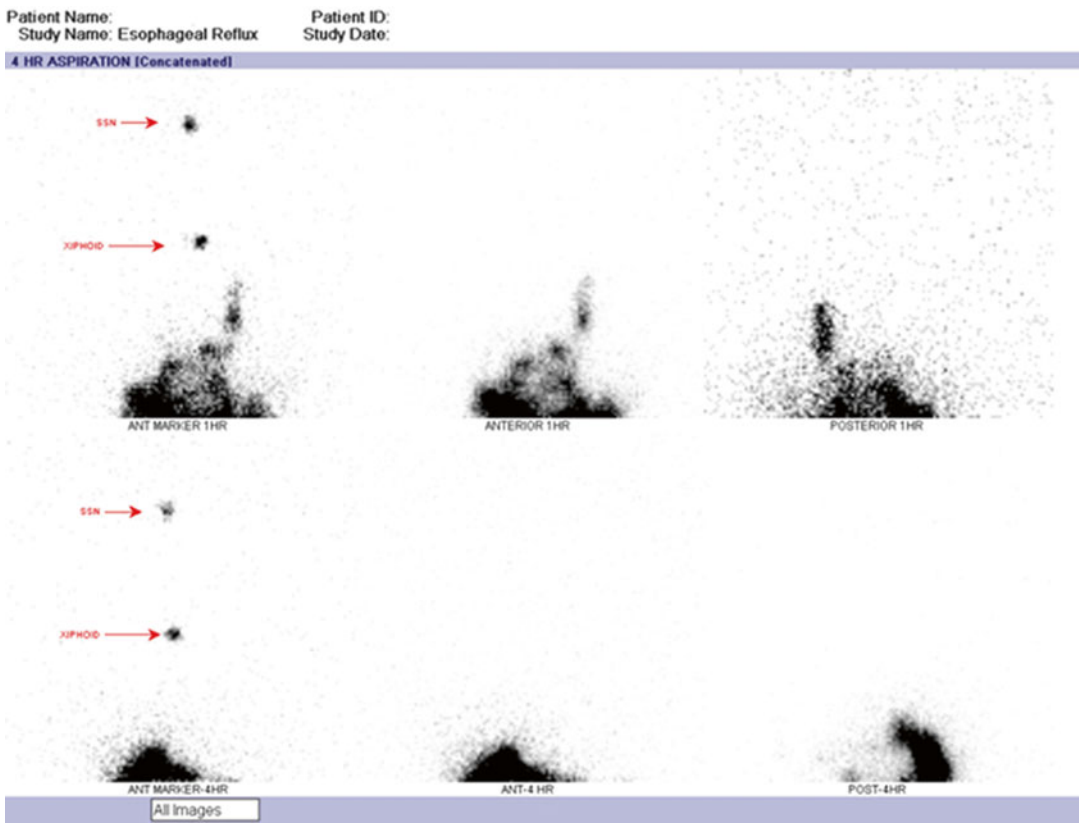

Fig. 39 Gastroesophageal reflux. Tc99m gastric emptying (radionuclide milk) scan showing gastric reflux to the level of the upper esophagus. Delayed imaging at 4 h reveals no pulmonary aspiration

episode and confirms pulmonary aspiration (Fig. 39). Practice guidelines for pediatric gastric scintigraphy have recently been updated and describe current techniques in more detail (ACR 2014). Scintigraphy is still undertaken in some centers to document response to treatment and stratify those who merit surgical intervention.

Passive reflux into the distal esophagus can be demonstrated sonographically (Koumanidou et al. 2004). The technique is performed after a suitable fast of approximately 2–4 h. Initial assessment, especially if there is no correlative barium study, concentrates on excluding gastric outflow tract obstruction including hypertrophic pyloric stenosis. According to some authors malrotation may also be assessed sonographically (Yousefzadeh 2009). Assessment of both these conditions is hampered when there is excessive aerophagia from crying in the starving child and a liquid feed should be given up to the normal amount. Using an age-appropriate curvilinear, or high-resolution linear probe with virtual trapezoid mode enabled, imaging is obtained with gentle graded compression in the transverse oblique plane with approximately 45°cranial tilt. The intra-abdominal esophagus is assessed as it enters through the diaphragm to the gastric cardia. Intra-abdominal esophageal length and transmural wall thickness including the serosa as well as the gastroesophageal angle of "His" are recorded. The number of episodes of reflux is also recorded. Color Doppler may be used to determine flow direction of moving content across the GEJ. Approximate study time is of the order of 15 min. Savino et al. (2012) have summarized normal values in children for mean esophageal

transmural wall thickness of 2–5 mm, esophageal diameter of 10 mm, esophageal length of 20–25 mm, and gastroesophageal angle of 70–100°. A short intra-abdominal esophageal length and more obtuse gastroesophageal angle correlate with GER. Gastroesophageal angle varies with gastric distension. Esophageal wall thickness is a nonspecific marker of GERD. Technical limitations of gastroesophageal US include failure to accurately document the cephalad extent of reflux and incomplete assessment of the thoracic esophagus which is largely obscured by artifact from the lungs. Currently there is no consensus on the sonographic criteria for an abnormal GEJ. Routine use of sonography is not recommended (Vandenplas et al. 2009) but is advocated in some centers.

5.2 Achalasia

Achalasia is an esophageal motility disorder characterized by failure of lower esophageal sphincter (LES) relaxation at the gastric cardia and absence of esophageal peristalsis with normal pharyngeal and upper esophageal sphincter function. This results from degeneration of the inhibitory myenteric plexus at the LES. Achalasia is rare with an annual estimated incidence of approximately 1/1000,000 in children (Lee et al. 2010). The most common symptoms are vomiting, dysphagia, regurgitation, and weight loss. In the neonatal period presentations are similar to those of GERD. Achalasia has been associated with trisomy 21, eosinophilic esophagitis, glucocorticoid insufficiency, congenital hypoventilation syndrome, familial dysautonomia, Chagas disease, and AAA (achalasia, alacrima, and ACTH insensitivity) syndrome (Hallal et al. 2012).

Definitive diagnosis is made with (barium) swallow study and esophageal manometry.

Unlike in adults endoscopic biopsy of the distal esophagus is not indicated, as the risk of associated malignancy is very low. Medical therapy includes endoscopic botulinum toxin injection and dilatation but affords only temporary symptomatic relief. Definitive surgical treatment in children with achalasia is laparoscopic Heller myotomy with or without a fundoplication procedure. However, the underlying esophageal dysmotility will persist and lifelong clinical follow-up is required.

5.2.1 Radiological Findings

The chest radiograph may reveal a dilated esophagus with an air-fluid level, changes of chronic pulmonary aspiration, and tracheal displacement (Fig. 40a, b). The gastric bubble is not visualized in cases with high-grade obstruction. A barium or water-soluble esophagram is the initial diagnostic study of choice; retention of a small oral barium tablet may reveal subtle disorders of motility. This may be followed by esophageal manometry and endoscopy. Contrast studies reveal a dilated esophagus that smoothly tapers distally to a "bird's beak" (Fig. 40c). Chronic untreated achalasia may lead to marked distension of the esophagus (Fig. 41). Rarely, a leiomyoma (see Sect. 5.10) may mimic the symptoms and radiological appearance of achalasia (Hussain et al. 2002).

5.3 Foreign Body Ingestion

The vast majority of foreign body ingestions occur in the pediatric population with a peak incidence between the ages of 6 months and 6 years. Esophageal foreign bodies tend to lodge at the normal anatomic sites of narrowing. In the upper GI tract, these include the cricopharyngeus muscle, GEJ, aortic arch, and the crossing of the left mainstem bronchus. Objects which pass beyond the cricopharyngeus will typically pass freely into the stomach though persistent lodging of a foreign body in the esophagus beyond this level is usually indicative of esophageal pathology such as a vascular ring or stricture from prior esophageal repair (Fig. 42). In order of frequency, esophageal foreign bodies lodge at the level of the thoracic inlet (53 %), thoracic esophagus (32 %), and the cervical esophagus (15 %) (Harned et al. 1997).

Children typically present with dysphagia and chest pain. Salivation and drooling occur with

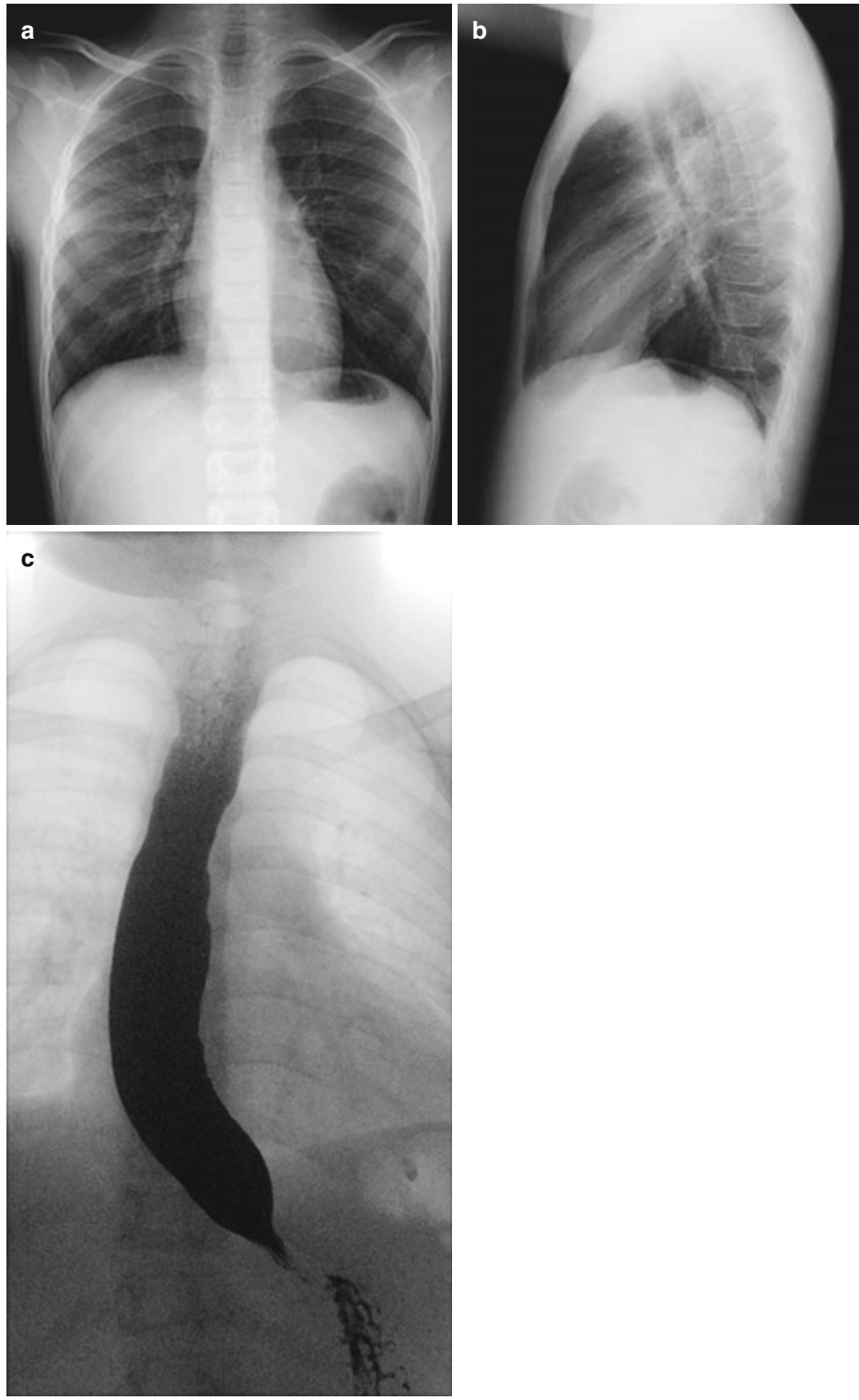

Fig. 40 (**a**–**c**) Achalasia. (**a**) AP and (**b**) lateral chest radiographs reveal an air-fluid level in the distended esophagus. (**c**) Barium esophagram demonstrating a patulous and dilated esophagus with distal *beaking* deformity

esophageal obstruction. Swallowed objects may be partially chewed food (Fig. 43) or other foreign bodies, two-thirds of which are coins in children (Webb 1995). Occasionally more than one object may be ingested. Acute coin ingestion is rarely symptomatic unless the coin is above the thoracic inlet (Sharieff et al. 2003). The majority of asymptomatic esophageal coins will pass spontaneously. Blunt objects which are well tolerated, with the exception of lithium disk batteries (see below), can be followed for 16–24 h (Sharieff et al. 2003; Waltzman et al. 2005). A lodged radiopaque esophageal foreign body should be removed if repeat radiography reveals non-passage. Flexible endoscopy enables foreign body extraction and provides immediate information about state of the esophagus at the site of impaction.

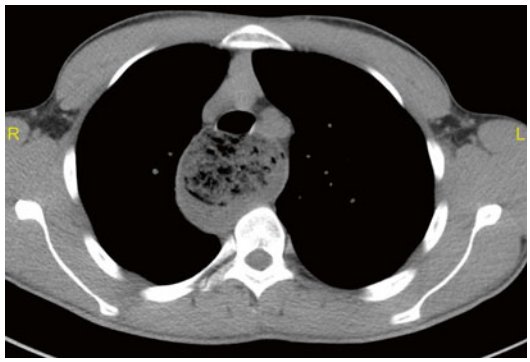

Fig. 41 Achalasia. Axial CT showing marked distension of the upper thoracic esophagus with retained food debris

5.3.1 Radiological Findings

Children are not always reliable historians. If there is concern for an ingested foreign body, then a lateral soft tissue neck radiograph and frontal radiograph from the oropharynx to the pubic symphysis (mouth-to-anus view) should be obtained. Radiographic evaluation of foreign bodies in a single center pediatric case series revealed 100 % of metal objects, 86 % of glass, and only 26 % of fish bones (Cheng and Tam 1999). Commonly ingested foreign bodies such as organic matter, medication, or small plastic objects are not visible on plain radiography. Radiolucent foreign bodies may show on the bar-

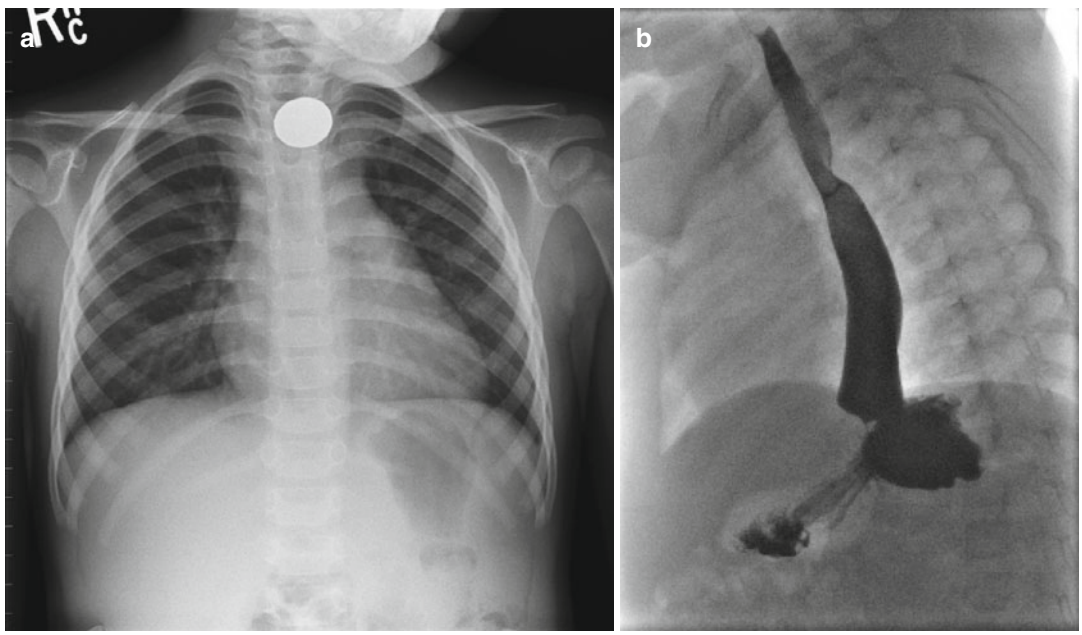

Fig. 42 (**a**, **b**) Impacted foreign body above an unsuspected vascular ring. (**a**) Frontal chest radiograph shows a coin lodged in the esophagus proximal to the aortic arch. (**b**) Barium esophagram confirms an oblique posterior indentation upon the esophagus in keeping a left-sided aortic arch and aberrant right subclavian artery

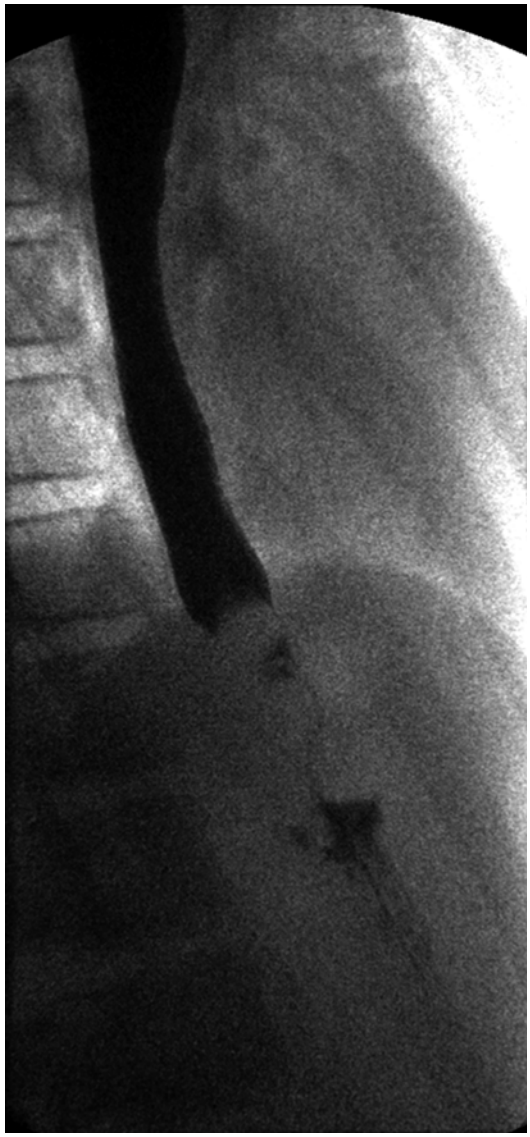

Fig. 43 Obstructing foreign body. Barium esophagram reveals a filling defect impacted in the distal esophagus. This was a beef steak morsel and ultimately required endoscopic removal

ium esophagram as filling defects in the barium (Fig. 43). Contrast studies should be avoided with a high-grade obstruction because of the risk of aspiration and may make endoscopic retrieval more challenging. Chronically impacted foreign bodies cause inflammation, edema of the esophageal wall, and lumenal narrowing. Complications of foreign body ingestion include perforation and abscess formation. An otorhinolaryngology (ENT) opinion is recommended for foreign bodies at or above the level of the cricopharyngeus. Emergent removal of esophageal impacted food boluses and a foreign body with evidence of complete esophageal obstruction is necessary. Multiple small neodymium magnets found in toys may inadvertently be ingested and lodge in the hypopharynx or esophagus. These can rapidly cause mucosal pressure necrosis and merit emergent endoscopic management (Brown et al. 2014). Surgical consultation is required for non-progression through the GI tract.

Alkaline lithium disk batteries (Fig. 44) contain very high concentrations of potassium or sodium hydroxide. Mucosal damage due to leakage occurs within 1 h of ingestion. Perforation may ensue within 8–12 h of esophageal impaction. This scenario is a medical emergency and mandates immediate removal. Failure to act promptly may result in the impacted battery eroding through the esophagus and into the airway or worse gives rise to an aortoesophageal fistula resulting in a life-threatening sentinel bleed (Brumbaugh et al. 2011). Esophageal bleeding should raise the suspicion of an ingested foreign body in a child.

5.4 Caustic Injury

Ingestion of both alkali and acidic agents may cause significant trauma to the mucosa of the esophagus with concomitant injury to the lips, mouth, oropharynx, and upper airway. The ensuing damage can include superficial mucosal burns and deep ulceration, progressing to full thickness injury. Liquefactive necrosis with fat saponification, protein denaturation, and thrombosis is seen with alkali agents. The mid and distal thirds of the esophagus are most vulnerable to alkaline agents and typically the stomach is spared. Conversely acidic agents are usually liquids and pass through the esophagus rapidly

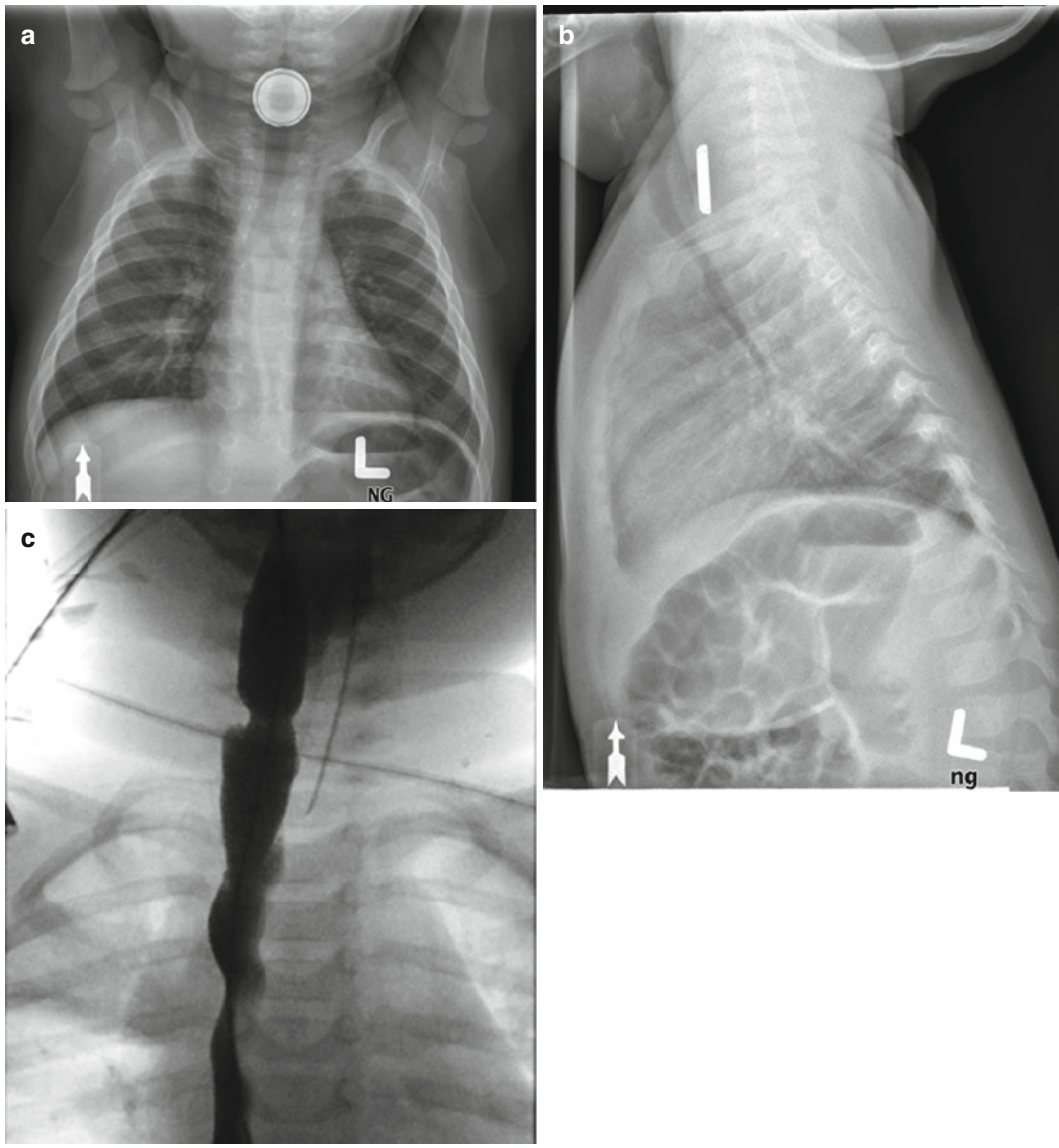

Fig. 44 (**a**–**c**) Lithium battery ingestion. (**a**, **b**) Frontal and lateral chest radiographs showing a retained lithium button battery in the hypopharynx/proximal cervical esophagus. (**c**) AP esophagram showing an irregular proximal esophageal stricture secondary to prior lithium battery ingestion

and tend to preferentially injure the stomach more severely. The risk of esophageal injury depends on the type of agent, form, amount, concentration, and duration of exposure. Accidental injury with alkaline bleaching (lye) agents typically occurs in small children aged 1–3 years where relatively small quantities are consumed. Rarely this may represent non-accidental injury in this age group. At the other end of the spectrum, ingestion of large quantities is seen in cases of deliberate self-harm in the adolescent population. Signs and symptoms of caustic agent ingestion are typically nonspecific. They may include coughing,

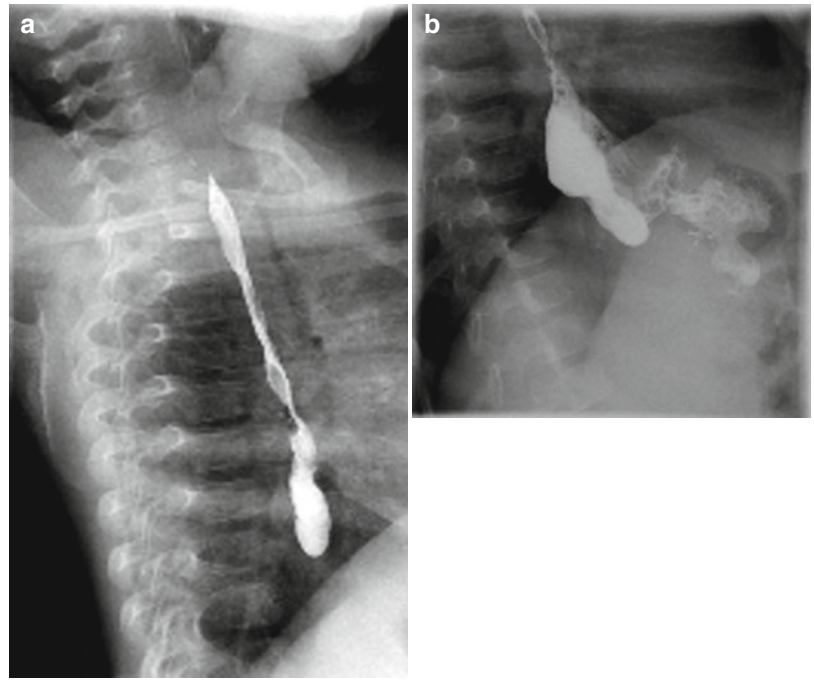

Fig. 45 (**a**, **b**) Caustic stricture. (**a**) Barium swallow showing an irregular high-grade stricture affecting the mid and distal thirds of the esophagus secondary to ingestion of an unknown caustic agent. The stomach is relatively spared. (**b**) Follow-up study showing an acquired hiatus hernia due to esophageal shortening

dyspnea, stridor, dysphonia, emesis, dysphagia, odynophagia, drooling, and hematemesis. Endoscopy is mandated in symptomatic patients (Betalli et al. 2008). The presence of retrosternal chest discomfort with back pain may indicate mediastinitis, while epigastric pain with rigidity may reflect peritonitis. Initial treatment commences with airway management and fluid resuscitation. Sips of water are encouraged to dilute and displace the chemical ingestant. Neutralizing agents and emetics are contraindicated. Surgery is rarely required.

5.4.1 Radiological Findings

Chest and abdominal radiographs are obtained to look for signs of perforation. A water-soluble contrast series is the initial radiological investigation of choice. Signs of injury include epiglottic edema, mucosal edema and ulceration, and esophageal dysmotility (Fig. 45). Intramural contrast tracking and persistent gaseous dilatation of the esophagus reflect severe injury and may precede perforation (Fernbach 1994). CT is recommended when there is a strong suspicion for perforation. Deep esophageal burns are investigated by serial barium esophagrams to detect early stricture formation. These are typically elongated and follow a protracted clinical course requiring periodic dilatations to provide adequate symptomatic relief. Where conservative therapy and serial pneumatic dilatation do not provide adequate palliation, surgery with colonic interposition (Fig. 46) or gastric pull-up is performed.

5.5 Esophageal Strictures

The most common cause for an acquired esophageal stricture in children is the result of a surgical repair of EA and TEF (Fig. 26). Other causes include caustic agent ingestion (Fig. 45), reflux esophagitis (Fig. 38), and epidermolysis bullosa (see Sect. 5.7). Symptoms relate to the degree of stricture. These include dysphagia, chest pain, coughing, vomiting of retained liquid or food, and aspiration pneumonia. Refusal to eat and drooling may indicate complete obstruction of the stricture by a retained foreign body.

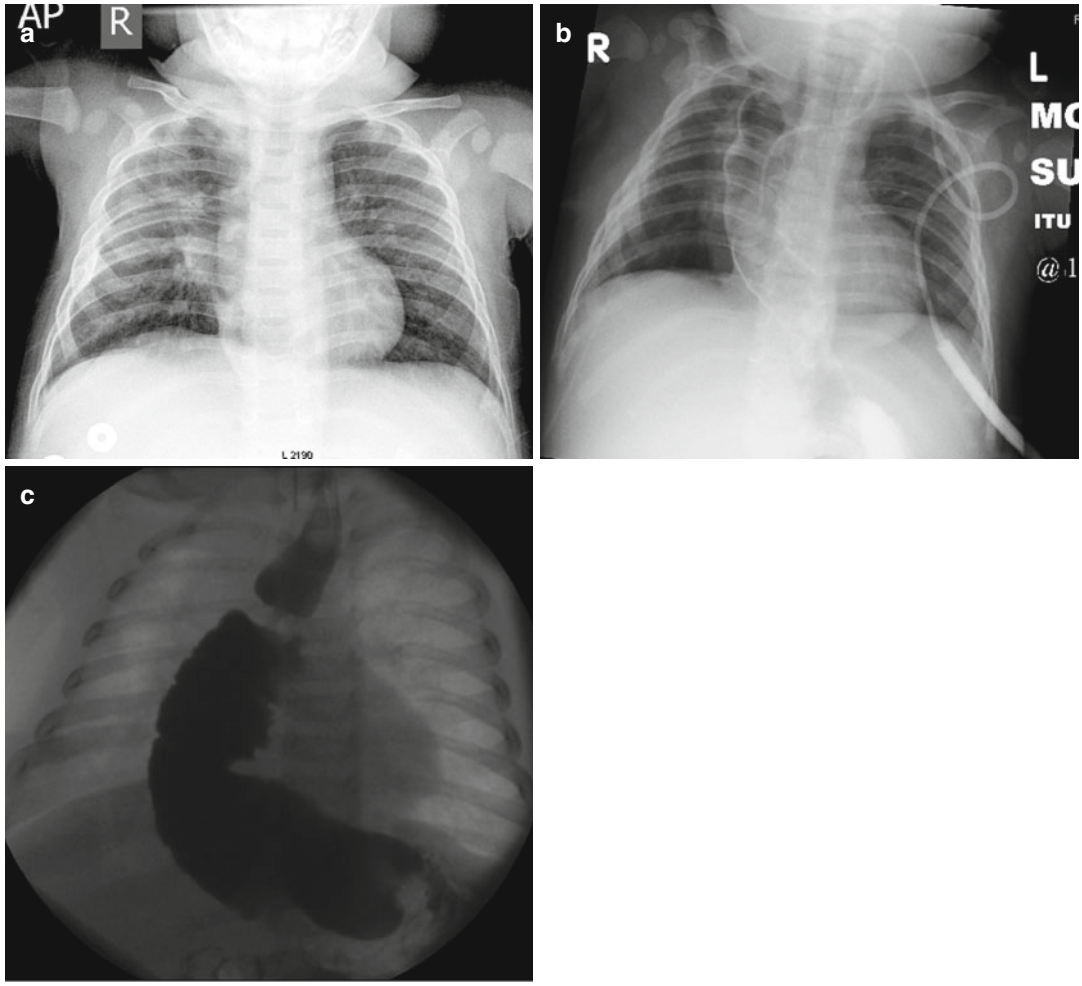

Fig. 46 (**a**–**c**) Esophageal interposition graft. (**a**) AP chest radiograph showing a right paravertebral posterior mediastinal air-filled lucent structure, (**b**) correlative barium study showing the colonic interposition graft. (**c**) Barium swallow in another child with a capacious colonic esophageal interposition graft

5.5.1 Radiological Findings

Radiographs are usually normal, although rarely an intraesophageal air-fluid level may be seen above an esophageal obstruction on an erect chest x-ray. A chest x-ray is however mandated if the patient's symptoms deteriorate following endoscopic treatment of a structure. A (barium) esophagram is the most appropriate study. Non-gastrografin water-soluble contrast medium may initially be used if a leak is suspected. Contrast studies reveal strictures as localized or diffuse regions of esophageal lumen narrowing with lack of distensibility (Fig. 47a) (Karasick and Lev-Toaff 1995).

Fluoroscopically guided balloon dilatation is the preferred treatment for esophageal stricture (Fig. 47b). Bougienage is an alternative but is limited by the diameter of the nares or pharynx. The incidence of perforation with balloon dilatation is also much lower. Balloon insufflation applies uniform radial force that is less traumatic than the shearing force of bougienage at the level of a stricture (Fasulakis and Andronikou 2003). Fibrotic scar tissue with its altered blood supply impairs tissue elasticity. As a result, strictures will typically not resolve after balloon dilatation but treatment does provide adequate temporary

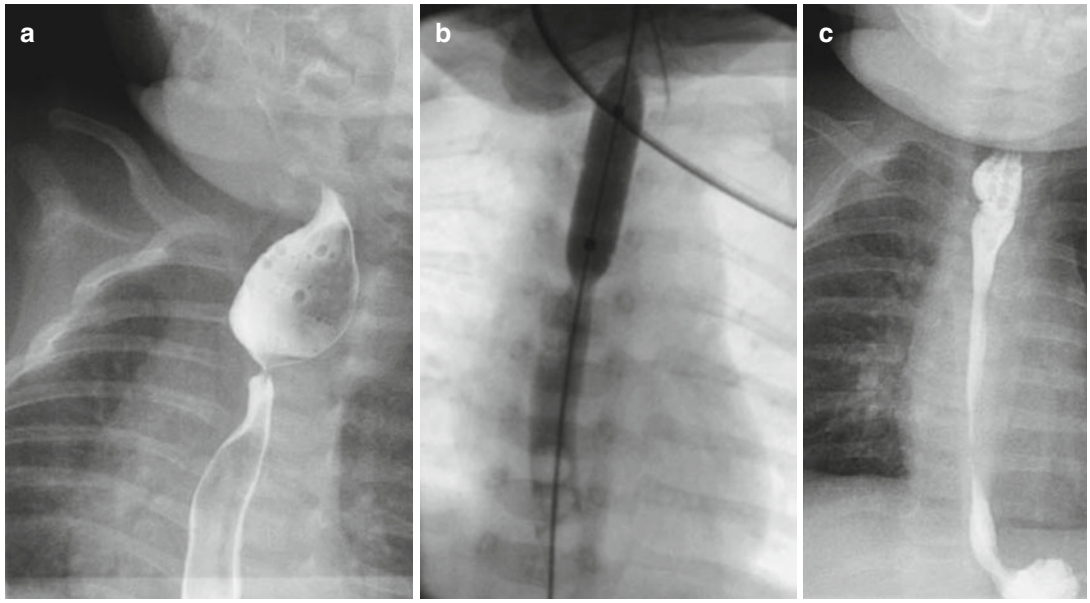

Fig. 47 (**a**–**c**) Balloon dilation of an esophageal stricture. (**a**) Severe stenosis of the proximal esophagus 3 months post-surgical repair. (**b**) Dilatation balloon expanded across the narrowing, no extravasation. (**c**) Post-dilatation appearance

symptomatic relief (Fig. 47c). Interval balloon dilatations are indicated in repaired esophageal atresia, caustic stricture, and epidermolysis bullosa. Serial balloon dilatations aid progressive stretching of scar tissue and reduce the risk of iatrogenic tears or perforation. Fluoroscopy also allows the radiologist to check that the stricture is dilated to a suitable diameter. A post-procedural water-soluble contrast study is performed to assess esophageal caliber and to evaluate for a leak and state of the distal esophagus. Recalcitrant esophageal strictures have been successfully treated with intralesional triamcinolone acetonide steroid (Lévesque et al. 2013) and topical mitomycin-C (Heran et al. 2008). Commercially available removable pediatric covered esophageal stents have been used albeit rarely for refractory benign esophageal strictures (Kramer and Quiros 2010). Surgery is rarely indicated for complex or refractory strictures.

5.6 Esophageal Perforation

Esophageal perforation is rare in the pediatric population. Iatrogenic esophageal perforation is the cause in 33–75 % of cases (Martinez et al. 2003). The most frequent sources of iatrogenic injury in infants and children include nasogastric tube insertion (Fig. 5), stricture dilatation, and trauma related to inadvertent esophageal intubation with an endotracheal tube (Gander et al. 2009). The incidence of iatrogenic perforation is rising as more diagnostic and therapeutic endoscopies are performed. The incidence is low in upper gastrointestinal endoscopy and higher with use of rigid dilators. Esophageal perforations are more likely to occur if an intraluminal foreign body has been present more than 24 h and result from sustained pressure necrosis. Other etiologies include caustic agents, pill-induced damage, and infection (including candida, herpes, and tuberculosis). Spontaneous rupture of the esophagus following vomiting or Boerhaave syndrome is rare in children (Fig. 48).

Symptoms relate to the level of perforation. Pharyngoesophageal and cervical esophageal perforation may result from peroral penetrating trauma including lollipops and pencils; symptoms may include neck pain and drooling. Symptoms of thoracic perforation include chest pain, vomiting, and crepitus related to subcutaneous

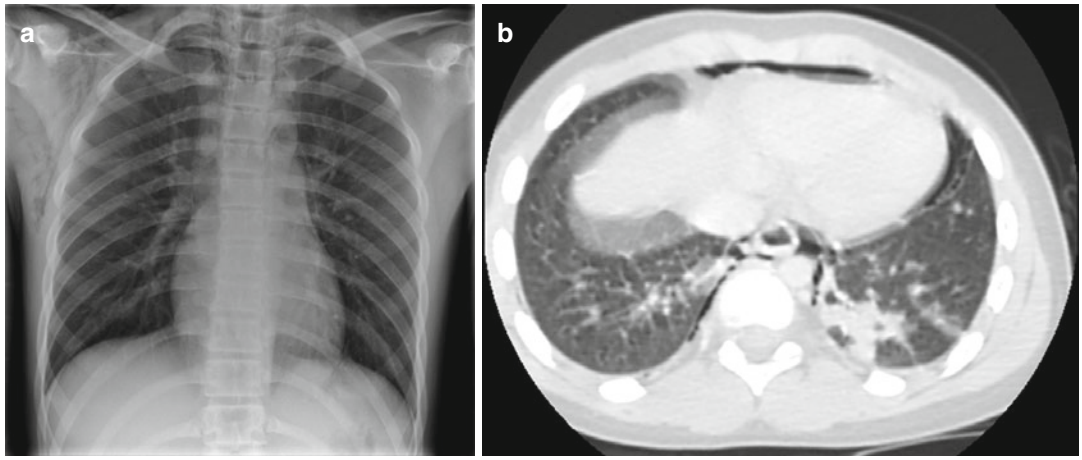

Fig. 48 (**a**, **b**) Esophageal perforation. (**a**) Frontal chest radiograph showing a pneumomediastinum with subcutaneous emphysema in the root of the neck, axillae, and supraclavicular fossa in a teenager following a bout of forceful vomiting. (**b**) Corresponding axial contrast-enhanced CT of the thorax

emphysema also known as the Mackler triad. Thoracic esophageal perforation is associated high morbidity and if untreated mortality because it allows direct entry of gastrointestinal flora and digestive enzymes to soil the mediastinum, pleural and subphrenic spaces causing sepsis. Intra-abdominal esophageal perforation may lead to sepsis, shock and pneumoperitoneum. Treatment for esophageal perforation is preferably conservative and non-operative. If there is no evidence of contrast leak at esophagram therapy consists of broad-spectrum antibiotic coverage, drainage of pleural effusions, esophageal rest and total parenteral nutrition. Successful outcome depends on early diagnosis and treatment, young age and absence of underlying disease (Martinez et al. 2003). Operative treatment may be required in hemodynamically labile patients to manage gross extraluminal contamination. Direct surgical repair to maintain luminal patency in large perforations is rarely indicated (Gander et al. 2009).

5.6.1 Radiological Findings

In cervical esophageal perforation airway-soft tissue neck radiographs are required as chest radiography is typically non-contributory. Chest radiographic findings in thoracic esophageal perforation include pneumomediastinum, subcutaneous emphysema, pneumothorax, hydropneumothorax, and pleural effusions (Fig. 48). The false negative rate with contrast swallows is around 10 % (Gimenez et al. 2002). CT is recommended when the esophagram is negative and there is high suspicion of perforation or when a preoperative roadmap is required particularly for thoracic esophageal injury.

5.7 Epidermolysis Bullosa

Epidermolysis bullosa (EB) is a rare inherited disorder. It is characterized with mechanical fragility of the skin and mucous membranes with progressive development of recurrent blisters and non-healing open wounds as a consequence of minor trauma resulting in high nutritional demands. Recessive dystrophic epidermolysis bullosa (RDEB) is a rare severe autosomal recessive form resulting from abnormal production of type VII collagen secondary to mutation of the COL7A gene which presents in the neonatal period.

Overall, four major types of EB are recognized with multiple subtypes and related genetic mutations. The main types are epidermolytic type (EB simplex), junctional EB, a dermolytic type (dystrophic EB), and a mixed type (Kindler syndrome). The most common finding is skin blistering. Some types and subtypes of inherited EB may be at risk for developing specific gastrointestinal complications and typically present in

the first or second decade of life. Esophageal bullae, stricturing, scarring, and web formation are found predominantly in the junctional type. Other complications include poorly coordinated swallowing, dysphagia, esophageal perforation, dysmotility, vomiting of a cast, and hiatus hernia. GERD occurs mainly in the non-junctional types (Fine and Mellerio 2009). Scarring and fibrosis may result in esophageal shortening predisposing to GERD and secondary stricturing (Anderson et al. 2004). Esophageal squamous carcinoma is a recognized complication. Syndromic associations include pyloric, esophageal, and anal atresia (Cetinkurşun et al. 1995).

The mainstay of treating EB esophageal stricturing is repeat fluoroscopic dilatation (Demirogullari et al. 2001). Gastrostomy provides esophageal rest. Rarely resection and colonic interposition are indicated. Children with epidermolysis bullosa require gentle handling. Modifications of imaging and anesthetic and therapeutic techniques are required to prevent iatrogenic blistering. Use of soft padding, spontaneous oral intake of contrast medium, and avoidance of restraints and enteric intubation are necessary. Dilatation is ideally performed in the symptomatic child when the underlying disease is quiescent.

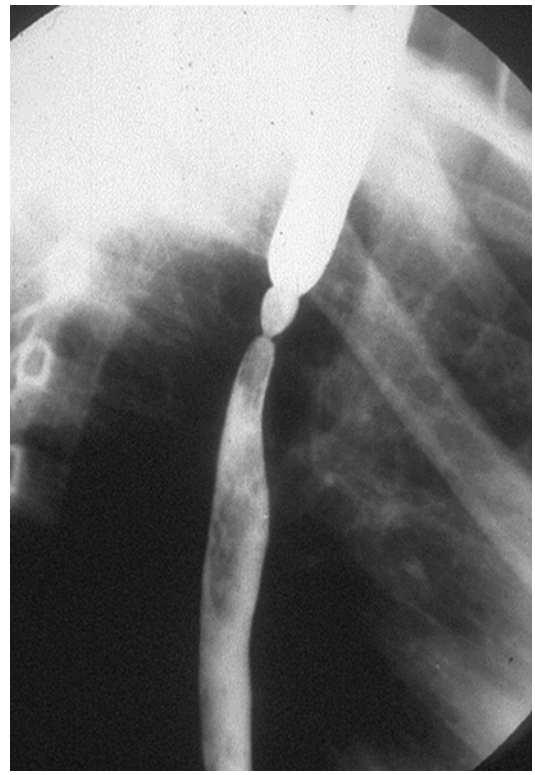

Fig. 49 Epidermolysis bullosa. Barium esophagram showing an annular proximal esophageal stricture in a child with the dystrophic, recessive form of the disease (Image courtesy of Dr. C Buonomo MD, Boston Children's Hospital, Boston, MA, USA)

5.7.1 Radiological Findings

A barium esophagram should evaluate the oropharynx and entire esophagus to demonstrate strictures and GERD. Approximately half of strictures are in the proximal third of the esophagus near the cricopharyngeus muscle (Fig. 49), a quarter in the distal one-third, and the remainder at multiple sites (Kern et al. 1989a). Strictures vary in length from several centimeters to annular strictures, which are typically less than 3 cm in length.

5.8 Infectious and Inflammatory Esophagitis

5.8.1 Infectious Esophagitis

Infectious esophagitis is rare in children. The most common cause is candida and usually occurs in immunocompromised patients as a complication of bone marrow transplantation, chemotherapy, human immunodeficiency virus (HIV), corticosteroid therapy, or rarely diabetes. Candida esophagitis may frequently occur in the absence of oropharyngeal candida (thrush) and should be suspected in the immunocompromised child presenting with odynophagia. Barium esophagram demonstrates shallow ovoid ulcers, discrete linear ulceration, and shaggy cobblestoned mucosal pattern. The differential for esophageal candidiasis includes herpes simplex virus (HSV) and cytomegalovirus (CMV), both of which give rise to similar imaging appearances but are refractory to antifungal therapy. HIV infection itself can produce large penetrating esophageal ulcers. HSV esophagitis under the age of 13 years is considered an HIV defining illness. Candida, CMV, and HSV esophagitis are HIV defining illnesses above 13 years in the absence of other risk factors (Baker 2013).

5.8.2 Eosinophilic Esophagitis

Eosinophilic esophagitis (EoE) is a chronic antigen-mediated disorder of unknown etiology manifesting as esophageal dysfunction with histological evidence of an eosinophil-predominant infiltrate (Liacouras et al. 2011). Although by definition, EoE is confined to the esophagus, EoE may be found in over 10 % of cases of atypical croup implying wider involvement of the aerodigestive tract (Cooper et al. 2012). EoE is often associated with atopic conditions such as asthma and allergies.

Symptoms include dysphagia, odynophagia, chest pain, food impaction, GERD-like symptoms refractory to medical and surgical therapy, abdominal pain, vomiting, early satiety, and anorexia (Liacouras et al. 2011). The prevalence and annual incidence of EoE in children has increased over time and is highest in children with food impaction or dysphagia (Soon et al. 2013). Treatment includes topical corticosteroids; systemic therapy is reserved for all but the most severe cases. The radiological manifestations of EoE (Fig. 50) have been described in a pediatric case series at a single institution (Binkovitz et al. 2010). In 24 UGI studies performed in 17 children with histologically proven EoE, 12 studies were normal, including four children with five episodes of recently treated acute esophageal food impaction. In the remainder long-segment nonulcerative strictures with mild mucosal irregularity were found in five cases, two in the mid-esophagus, one in the distal esophagus, and two at the GEJ. Only one stricture was demonstrated at endoscopy highlighting the usefulness of the UGI in delineating strictures. The finding of a Schatzki ring raises the possibility of EoE (Fig. 51). A Schatzki ring is a symptomatic, intermittent or persistent, discrete circumferential submucosal narrowing which distorts the esophageal lumen at the level of the gastroesophageal junction. The Schatzki ring differs from an asymptomatic esophageal B-ring found up to 1 cm cephalad to the esophageal diaphragmatic hiatus. In a single center retrospective study of UGI series in a pediatric and adolescent population, the incidence of Schatzki ring was found to be 0.2 % and was associated with hiatus hernia, GERD, and EoE with the authors recommending endoscopic biopsy in all cases to exclude the latter (Towbin and Diniz 2012).

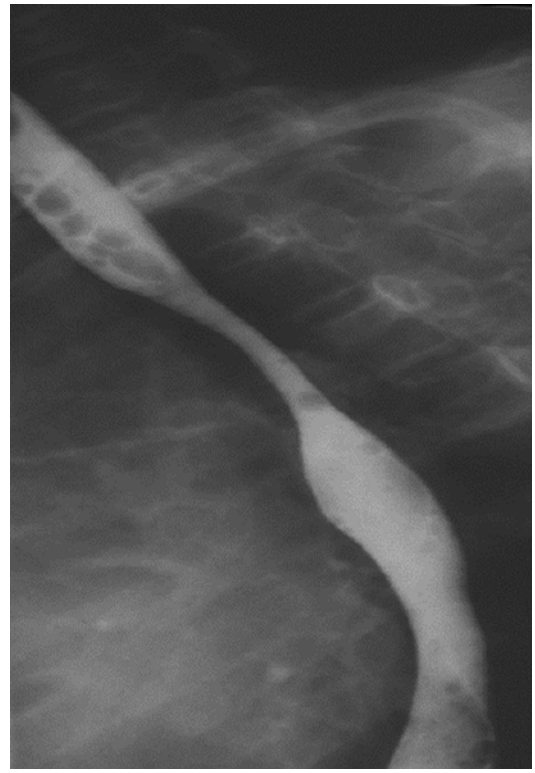

Fig. 50 Eosinophilic esophagitis. Barium esophagram revealing a proximal long-segment nonulcerative stricture with mild mucosal irregularity (Image courtesy of Dr. C Buonomo MD, Boston Children's Hospital, Boston, MA, USA)

5.8.3 Crohn Disease

Although Crohn disease can affect any portion of the alimentary tract, esophageal involvement is rare. Radiological appearances are similar to those of adults and include aphthous ulceration, filiform polyposis, deep ulceration, fissuring, sinus tract formation, fistulae, and stricturing (Fig. 52).

5.8.4 Graft Versus Host Disease

Graft versus host disease (GVHD) is a multisystem complication following bone marrow transplantation, more commonly seen in allogenic rather than autologous transplants. In acute esophageal GVHD

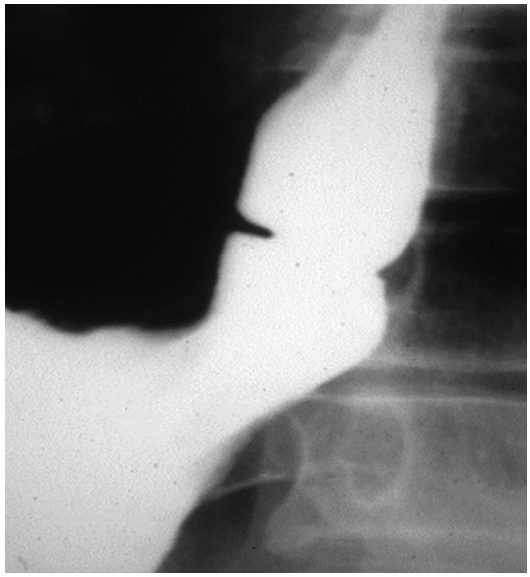

Fig. 51 Schatzki ring. Barium esophagram showing a symptomatic circumferential B-ring in the distal esophagus (Image courtesy of Dr. C Buonomo MD, Boston Children's Hospital, Boston, MA, USA)

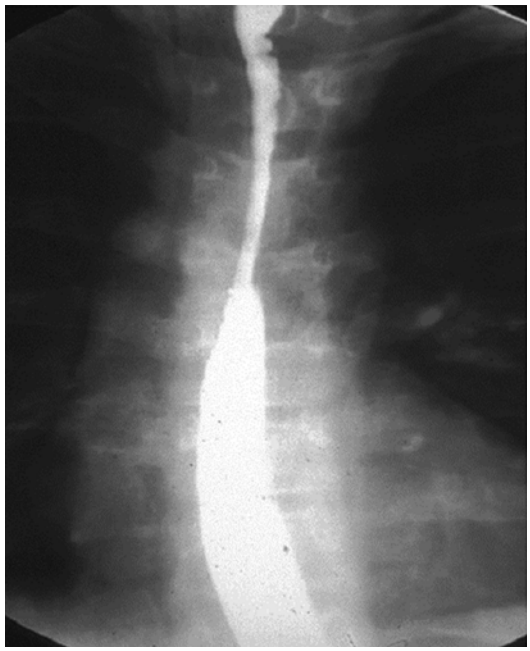

Fig. 52 Esophageal Crohn disease. Barium esophagram showing an irregular long-segment stricture in the proximal esophagus. Esophageal involvement is rare in Crohn disease (Image courtesy of Dr. C Buonomo MD, Boston Children's Hospital, Boston, MA, USA)

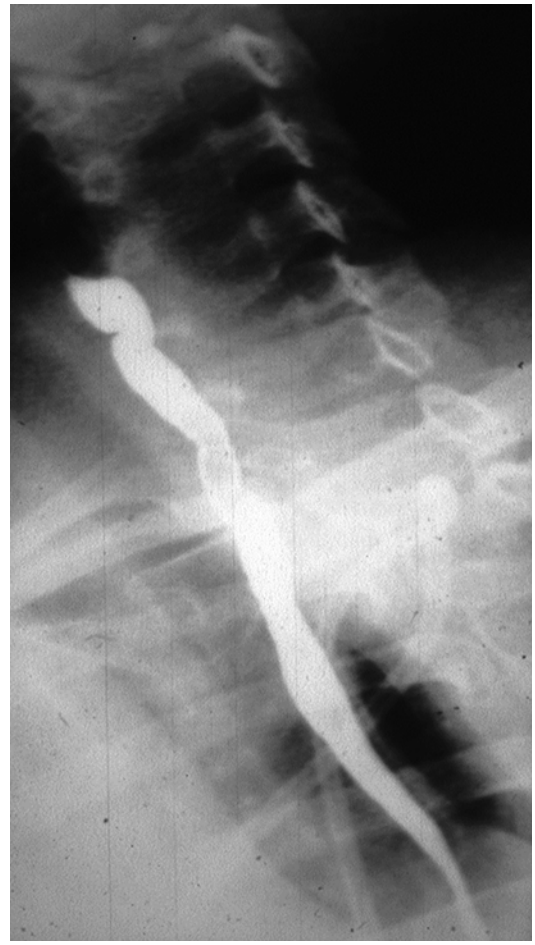

Fig. 53 Esophageal graft versus host disease. Barium esophagram showing a chronic mid-upper esophageal web formation and stricturing (Image courtesy of Dr. C Buonomo MD, Boston Children's Hospital, Boston, MA, USA)

donor T lymphocytes and cytokines mediate an inflammatory superficial mucosal esophagitis. Chronic GVHD results in patchy submucosal fibrosis and focal stricturing. Radiological appearances at UGI include aperistalsis, webs, circumferential narrowing, and proximal and mid esophageal tapered strictures (Fig. 53).

5.9 Esophageal Varices

Esophageal varices are less common in children than in adults. Varices are a late complication of chronic pediatric liver disease. They typically

occur in the setting of portal hypertension secondary to umbilical venous catheterization, biliary atresia, alpha-1 antitrypsin deficiency, autosomal recessive polycystic renal disease, and cystic fibrosis. Flow through the variceal esophageal collateral veins is typically hepatofugal or "upstream" and toward the superior vena cava. At endoscopy these typically affect the distal esophagus. Downhill esophageal varices are extremely rare in children and occur in the setting of gradual superior vena cava (SVC) occlusion and may be suspected at endoscopy when the entire length of the esophagus is affected in the absence of portal hypertension. Upper esophageal involvement only implies SVC obstruction proximal to the inflow of a patent azygos vein.

5.9.1 Radiological Findings

On (barium) swallow, the normal parallel mucosal folds are interspersed by serpiginous variceal filling defects that are best appreciated on collapsed views of the esophagus. Doppler ultrasound, dynamic MR angiography, and MR cholangiography have largely supplanted barium studies in the diagnostic work-up of these children. Doppler ultrasonography (US) is the first-line imaging technique in children with liver cirrhosis and suspected varices. Portal vein diameter and flow velocity yield the congestion index. Splenic size, abnormal hepatic vein flow patterns, and presence of abdominal portosystemic collaterals are markers of portal hypertension. US has limited specificity for detecting large esophageal varices; alternatives include wireless capsule endoscopy and conventional endoscopy, the latter enables therapeutic intervention. Transient elastography (TE) is a noninvasive sonographic technique used to evaluate fibrosis in chronic liver disease. TE has been shown to be a useful noninvasive surrogate marker for the presence of esophageal and gastric varices in children following portoenterostomy for congenital biliary atresia (Chongsrisawat et al. 2011). Correlation with variceal size and bleeding risk was not documented. MR elastography (MRe) is an alternative to TE in children. A single study has depicted gastroesophageal variceal formation in two cases at MRe (Binkovitz et al. 2012).

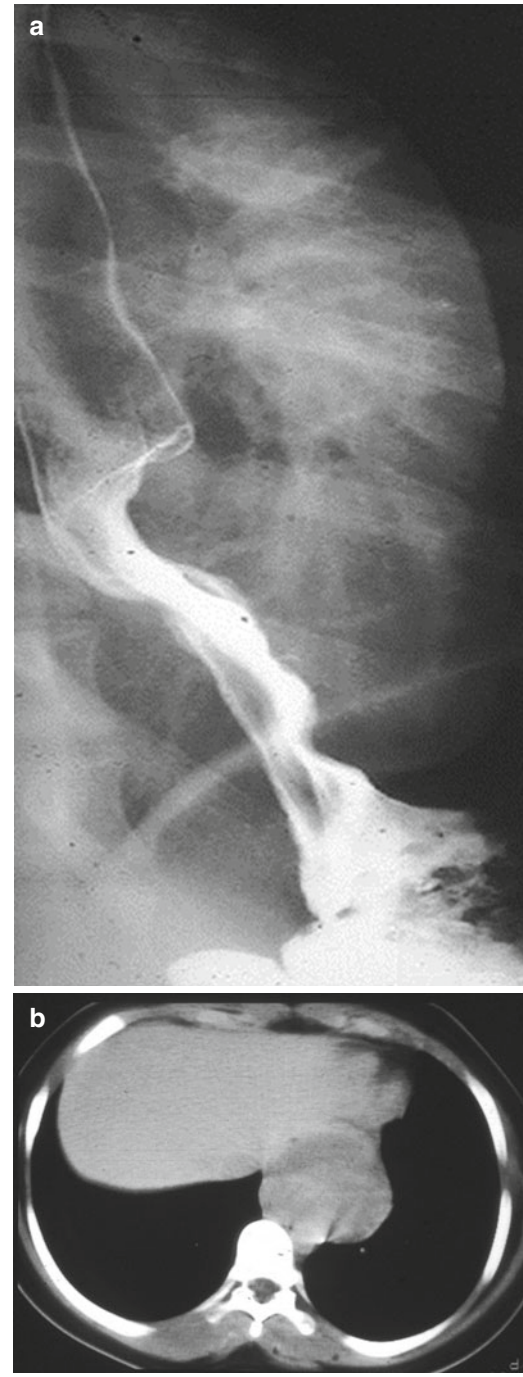

Fig. 54 (**a**, **b**) Esophageal leiomyoma. (**a**) Barium esophagram demonstrating a submucosal filling defect within the distal third of the esophagus. (**b**) Corresponding axial contrast-enhanced CT showing a heterogeneous soft tissue mass centered on the esophagus and azygoesophageal recess (Images courtesy of Dr. C Buonomo MD, Boston Children's Hospital, Boston, MA, USA)

5.10 Esophageal Neoplasms

Pediatric esophageal neoplasia is rare. Symptoms are nonspecific but include dysphagia, odynophagia, chest pain, and weight loss. Leiomyomas (Fig. 54) are the most common benign esophageal neoplasm and are typically seen in teenage girls. Diffuse esophageal involvement is seen in 90 % of cases; in less than 10 % patients, they may present with a focal mass (Hryhorczuk et al. 2013). Leiomyoma is associated with Alport syndrome and familial leiomyoma syndromes. Lymphoma may involve the pediatric esophagus but rarely so. Esophageal carcinoma is extremely rare. Risk factors for adenocarcinoma include caustic ingestion, reflux disease, and Barrett esophagus. Risk factors for squamous cell carcinoma include caustic ingestion, inherited bone marrow failure syndromes, and epidermolysis bullosa. Owing to the rarity of pediatric esophageal neoplasia, there are no consensus guidelines in the approach to management and the prognosis is very poor for solid malignancies (Issaivanan et al. 2012).

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