Pediatr Radiol (2014) 44:467–475
DOI 10.1007/s00247-013-2847-2

PICTORIAL ESSAY

Imaging of long gap esophageal atresia and the Foker process: expected findings and complications

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Received: 24 June 2013 / Revised: 13 October 2013 / Accepted: 21 November 2013 / Published online: 24 December 2013

Abstract Long gap esophageal atresia (EA) is characterized by esophageal segments that are too far apart for primary anastomosis. Surgical repair utilizing interposition grafts or gastric transposition are often employed. The Foker staged lengthening procedure is an alternative surgical method that utilizes continuous traction on the esophagus to induce esophageal growth and allow for primary esophageal anastomosis. This pictorial review presents the step-by-step radiographic evaluation of the Foker procedure and illustrates the radiographic findings in the most commonly encountered complications in our cohort of 38 patients managed with this procedure from January 2000 to June 2012.

Keywords Long gap esophageal atresia · Foker process · Children · Esophagus · Esophagram

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Introduction

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is a rare congenital disorder, occurring in approximately 1:4,500 live births [1]. The term long gap esophageal atresia is applied when the distance between the upper and lower atretic segments is too far for primary anastomosis. Because primary anastomosis is not possible in this group of patients, several surgical techniques are utilized to establish continuity between the atretic segments. These include 1) primary repair under tension; 2) gastric mobilization with partial gastric pull up; 3) lengthening myotomies; 4) proximal and distal pouch stretching by various methods, and 5) esophageal replacement with gastric, colon or jejunum interpositions [2–4]. In 1997, Foker et al. [5] described an alternative technique that utilizes traction sutures to promote in vivo esophageal growth through tension-induced lengthening and subsequent delayed primary repair. Therefore, the Foker process allows for the native esophagus to serve as the conduit, even when the atretic segments are widely spaced.

The Foker process is a two-stage surgical procedure. Stage I consists of placing the esophageal segments under traction (Fig. 1). Stage II is the esophageal anastomosis after traction-induced growth. As with all EA patients, there is a high incidence of gastroesophageal reflux (GER) in these children and a large number of patients subsequently go on to gastric fundoplication. It is important that radiologists are familiar with the expected radiographic findings at each stage as well

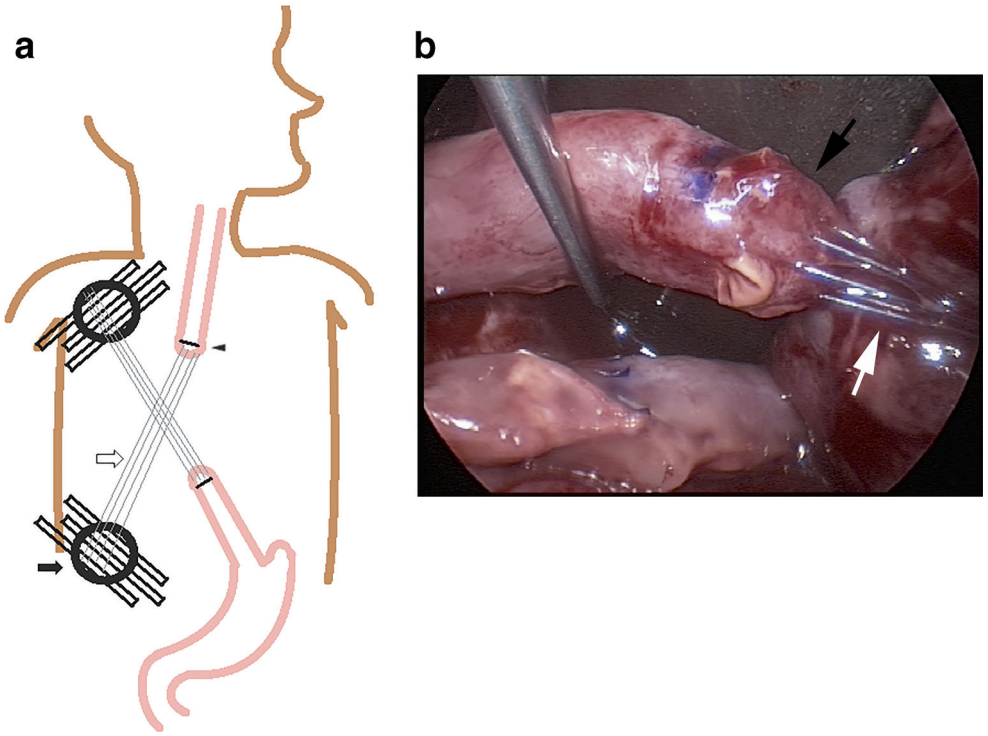

Fig. 1 Foker Stage I. **a** Schematic demonstrates esophageal segments marked by radiopaque clips (*black arrowhead*) and attached to lengthening apparatuses in the chest wall (*black arrow*) with traction sutures (*white arrow*). **b** Intraoperative photograph demonstrates an upper esophageal segment (*black arrow*) with traction sutures in place (*white arrow*)

as commonly encountered complications in this patient group [6–8]. This Institutional Review Board-approved pictorial essay will highlight the imaging findings encountered in our cohort of 38 children managed with this procedure from January 2000 to June 2012 and will outline what radiologists need to know.

Imaging prior to surgery

Plain radiographs are frequently the first imaging test obtained in this patient group. After birth, initial radiographs may demonstrate a dilated upper esophageal segment or a nasoenteric tube terminating in the upper esophageal segment. Eighty-five percent will have a distal TEF and air in the GI tract and 5% will have pure EA with no fistula and the abdomen will be gasless (Fig. 2). EA is associated with additional anomalies in approximately 50% of cases, the majority involving one or more of the VACTERL association (vertebral, anorectal, cardiac, tracheoesophageal, radial ray/renal and limb anomalies) [9]. Therefore, particular attention should be paid to the presence of associated anomalies when interpreting radiographs of patients with EA.

Once the diagnosis of EA is established, the length of the gap is evaluated by performing a "gap-o-gram" esophagram, which requires access via a percutaneous gastrostomy. If a gastrostomy is present, patients are brought to the fluoroscopy suite and a nasoesophageal catheter is positioned with its tip in the proximal esophageal segment and a second catheter is positioned with its tip in the distal segment via a gastrostomy. The gap length is determined by injecting a water-soluble contrast agent such as ioversol (Optiray-300; Mallinckrodt, St. Louis, MO, USA) and referencing a calibrated ruler placed in the image field (Fig. 3). If there is reflux into the distal esophageal segment, positioning the distal catheter in the stomach is often sufficient. In our institution the surgical team is present during the esophagram and places the enteric tubes. We use 5-Fr catheters. If gastrostomy is not present, the initial "gap-o-gram" esophagram may be performed in the operating room at the time of gastrostomy placement.

Imaging after traction suture placement (Stage I)

Radiographic evaluation after traction suture placement depends largely on the clinical situation. All patients receive frequent chest radiographs. Additional studies may include follow-up esophagram, US and/or CT as clinically indicated. In the preoperative period, care must be taken when placing the enteric tubes

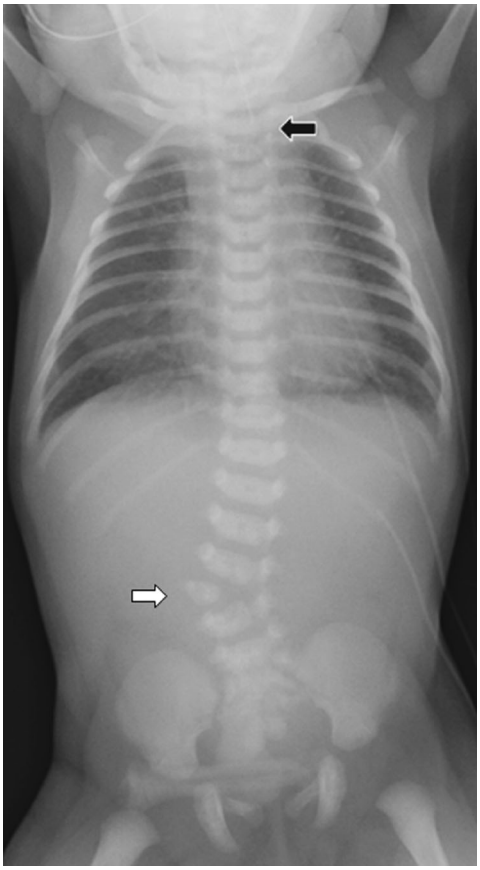

Fig. 2 Frontal radiograph of the chest and abdomen in a newborn male with esophageal atresia. The abdomen is gasless, a nasoenteric tube terminates in an upper esophageal pouch (*black arrow*) and a lumbar hemivertebra (*white arrow*) is noted

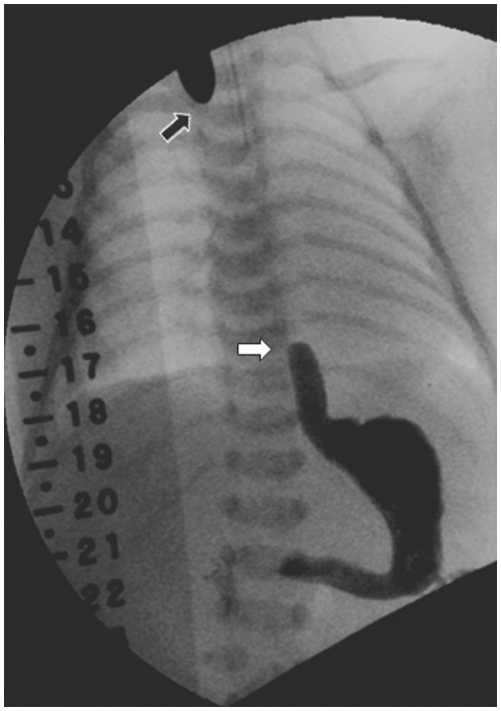

Fig. 3 Presurgical "gap-o-gram" esophagram in a 2-day-old boy with esophageal atresia. Intraoperative "gap-o-gram" esophagram with a calibrated ruler demonstrates a 5.5-cm gap between the proximal (*black arrow*) and distal (*white arrow*) esophageal segments

when performing "gap-o-gram" esophagram due to the fragility of the esophageal pouches and risk of iatrogenic perforation.

Expected findings after Stage I

Stage I of the Foker process involves thoracotomy, placement of surgical clips at the end of each esophageal segment to act as radiographic markers and placement of traction sutures that are externalized to lengthening apparatuses on the skin (Figs. 1 and 4). Tension is induced by periodically adding catheter tubing to the lengthening apparatuses. Esophageal growth is monitored on serial chest radiographs (Fig. 5) by tracking the position of clips and on "gap-o-gram" esophagrams (Fig. 6) by tracking the position of the opacified esophageal lumen.

Complications after Stage I

Complications occurring after Stage I may include esophageal segment leak, empyema and/or abscess. Patients are sedated

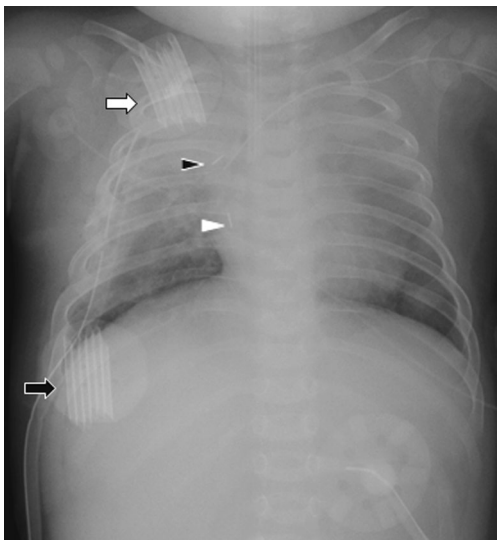

Fig. 4 Chest radiograph in a 6-month-old boy after Stage I. The proximal esophageal segment is marked by a clip (*black arrowhead*) and attached to the lower lengthening apparatus (*black arrow*). The distal esophageal segment is also marked by a clip (*white arrowhead*) and attached to the upper lengthening apparatus (*white arrow*)

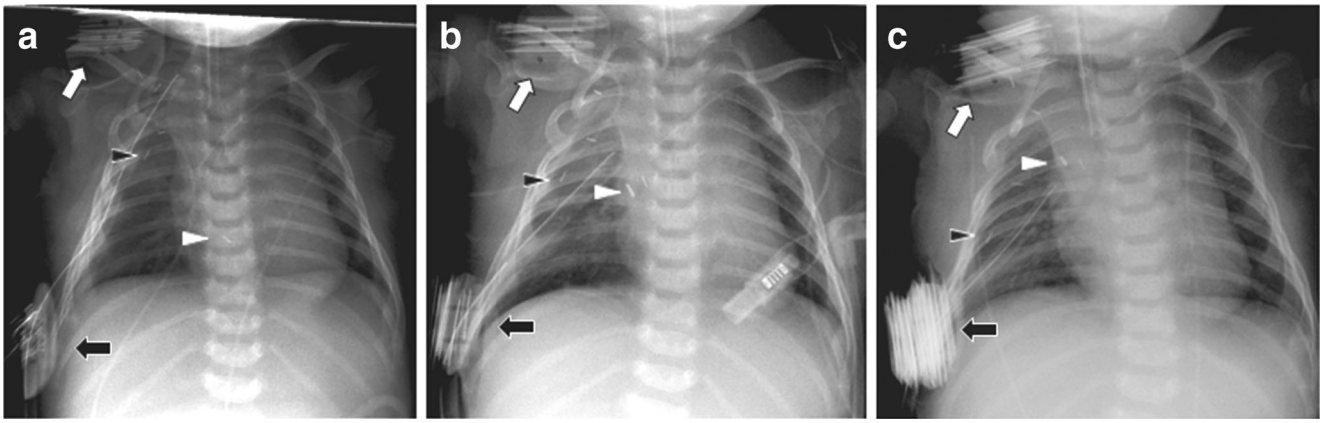

Fig. 5 Serial chest radiographs in a 1-month-old boy undergoing the Foker process demonstrate traction-induced esophageal growth. The ends of the esophageal segments are marked by clips (*arrowheads*) and the traction devices consist of anchors and pieces of catheter tubing (*arrows*). The proximal esophageal segment (*black arrowhead*) is attached to the lower device (*black arrow*) and the distal esophageal segment (*white arrowhead*) is attached to the upper device (*white arrow*). Chest radiographs on postoperative day 1 (**a**), 5 (**b**) and 11 (**c**) demonstrate increased catheter tubing, which provides continuous traction, and movement of the esophageal markers indicating tension-induced growth, with several centimeters of overlap by postoperative day 11. Subsequent "gap-o-gram" esophagram (not shown) demonstrated intact overlapping esophageal segments

and paralyzed throughout Stage I, but paralysis is periodically lifted and suture disruption may occur. If there is disruption of an esophageal segment, extraluminal gas and debris may be seen on radiographs and leak may be confirmed on "gap-o-gram" esophagram (Fig. 7). Infections of the chest cavity, often related to leak, can lead to empyema or abscess (Fig. 8).

Imaging after esophageal anastomosis (Stage II)

Expected findings after Stage II

Once adequate esophageal length is achieved patients undergo Stage II, consisting of repeat thoracotomy and esophageal anastomosis. After anastomosis, an esophagram

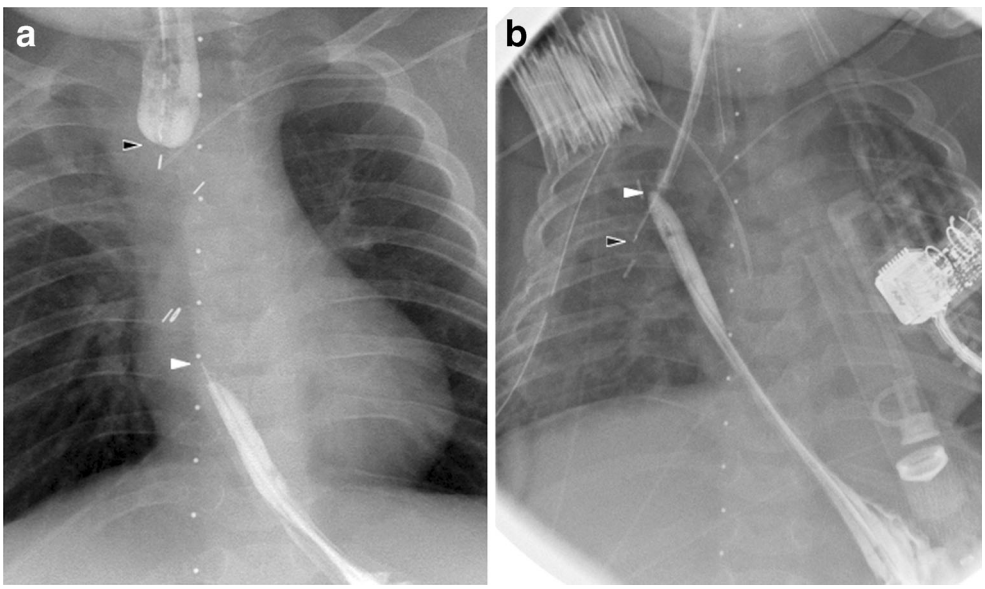

Fig. 6 "Gap-o-gram" esophagrams demonstrate tension-induced growth of the proximal (*black arrowhead*) and distal (*white arrowhead*) esophageal segments, occurring between esophagram performed on the day after transfer to our institution (**a**) and after 14 days of tension-induced growth (**b**)

Fig. 7 Pouch leak in a patient undergoing the Foker process. Contrast medium is injected through catheters positioned in the proximal and distal pouches, and leak (*arrow*) is seen arising from the proximal pouch

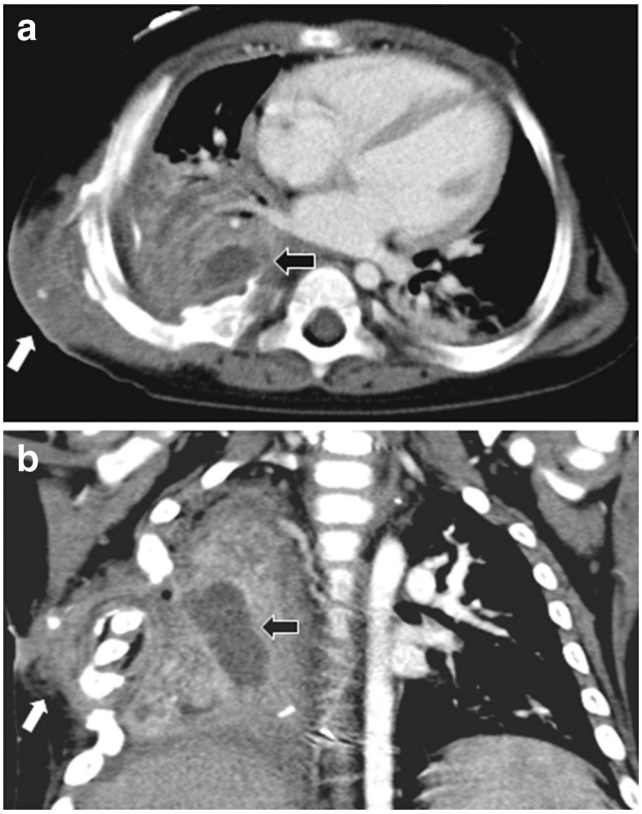

Fig. 8 Contrast-enhanced CT of the chest in a 2-year-old boy with long gap esophageal atresia undergoing the Foker process with known lower segment leak, fever and leukocytosis. Axial (**a**) and coronal (**b**) images demonstrate a peripherally enhancing empyema (*black arrows*) with extension of phlegmon to the chest wall (*white arrows*)

is performed by administering an oral feed or positioning a nasoenteric tube within the proximal esophagus and injecting water-soluble contrast medium under fluoroscopic observation (Fig. 9). Care must be taken to avoid disrupting the fragile anastomosis if performing the esophagram via a nasoenteric tube. In our institution, the surgical team is present during the esophagram and positions the nasoenteric tube for the study. Contour irregularity is often noted at the anastomotic site, but the esophagus is expected to be widely patent without leak.

Complications after Stage II

The most common complications following Stage II are esophageal stricture and leak. Surgically induced tension and GER are both thought to increase the risk for these complications [7, 10]. Strictures are often identified on esophagram and are treated with balloon dilatation (Fig. 10). Removable covered stents may be utilized in selected cases of recalcitrant stricture [11, 12] (Fig. 10). Leaks can be suggested by findings of new pleural fluid on chest radiograph or US and are confirmed on esophagram (Fig. 11). Leaks tend to occur at the anastomosis and occur over a broad time interval with later leaks often occurring after stricture dilation. Like pouch leaks, anastomotic leaks predispose to empyema and abscess. Additional findings after Stage II include gastroesophageal reflux (Fig. 12) and hiatal hernia (Fig. 13).

Imaging after gastric fundoplication

Expected findings after fundoplication

There is a high incidence of GER in all patients with EA, and when reflux occurs after anastomosis this may lead to

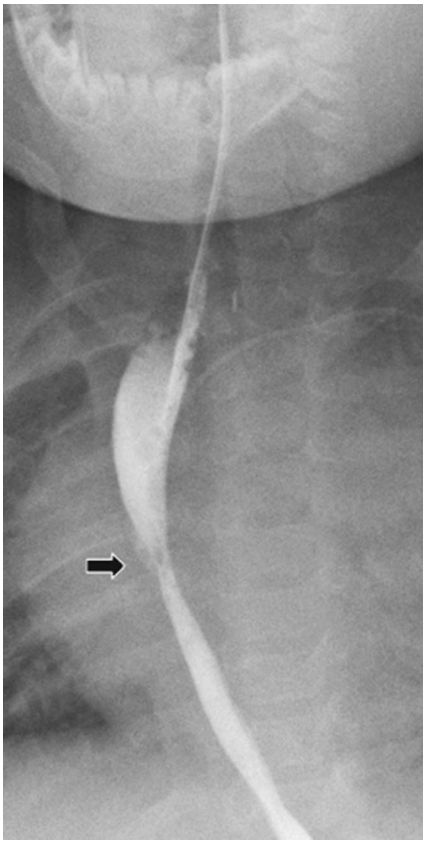

Fig. 9 Satisfactory postoperative esophagram in a 7-month-old girl, postoperative day 13 after anastomosis. Nasoenteric tube is positioned within the proximal esophagus and water-soluble contrast medium is injected. Mild narrowing (*arrow*) at anastomosis is expected in the immediate-postoperative period

an increased incidence of strictures and leaks [7, 10]. Gastric fundoplication is performed to help alleviate this. Imaging after fundoplication begins with gastrostomy injection to evaluate for gastroesophageal reflux. If no reflux is present, the patient swallows contrast medium and esophagram is performed. On esophagram, the fundoplication wrap is seen (Fig. 14) and there should be timely antegrade passage of contrast medium through the wrap without obstruction.

Complications after fundoplication

Complications after fundoplication include delayed transit across the fundoplication and persistent gastroesophageal reflux (Figs. 15 and 16). Delayed transit may be related to postoperative edema and resolve with time or due to a tight configuration of the wrap requiring balloon dilatation or surgical revision. Persistent gastroesophageal reflux often requires surgical revision.

Osseous findings: fracture and chest wall deformity

Patients undergoing the Foker process for the treatment of long gap EA have a high incidence of fractures. Fifty percent of patients in our cohort sustained a long bone fracture. Buckle-type and minimally displaced fractures are most common, and they most frequently occur in the proximal humerus and distal femur. Prolonged paralysis and fluid restriction lead to osseous demineralization and

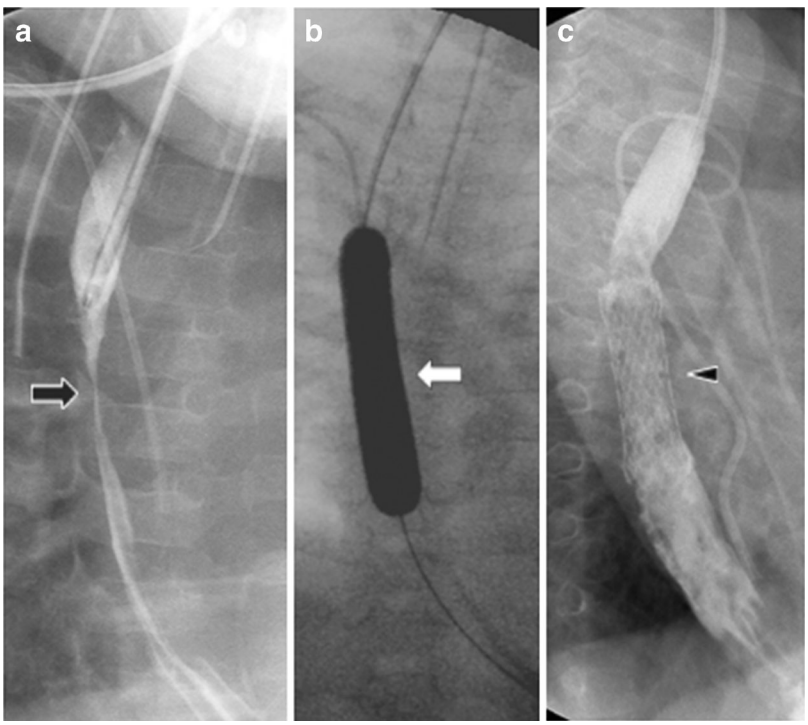

Fig. 10 Esophageal stricture, balloon dilatation and stent after the Foker process. Esophagram (**a**) on postoperative day 13 demonstrates mid-esophageal stricture (*black arrow*). Balloon dilatation (*white arrow*, **b**) was performed several times to treat the esophageal stricture. A persistent stricture was treated with a covered removable esophageal stent (*black arrowhead*). Esophagram (**c**) demonstrates patency of the esophagus through the stent

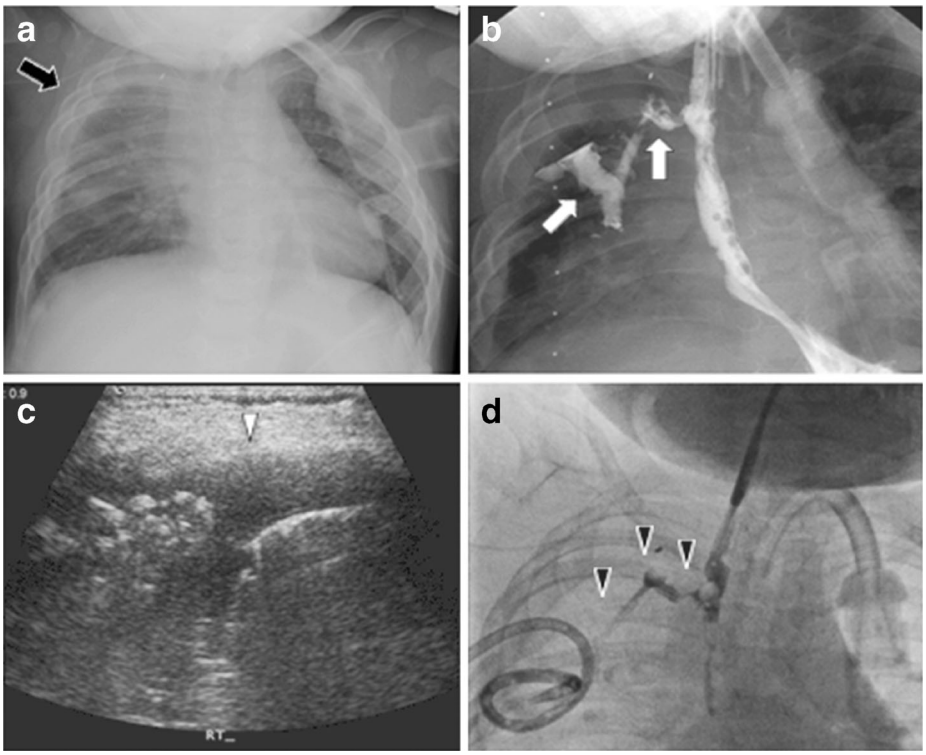

Fig. 11 A 15-month-old boy with long gap EA undergoing the Foker process, status post esophageal anastomosis with esophageal leak on postoperative day 18. Chest radiograph (**a**) demonstrates pleural fluid (*black arrow*). Esophagram (**b**) demonstrates a leak with esophagopleural fistula (*white arrows*). Ultrasound (**c**) demonstrates pleural fluid (*white arrowhead*). A 10-Fr pleural pigtail catheter was placed. Fluoroscopic image obtained after nasoenteric tube contrast injection (**d**) demonstrates persistent leak (*black arrowheads*)

Fig. 12 Gastrostomy port injection of a gastrojejunostomy tube in a 2-month-old boy 14 days after anastomosis demonstrates GER

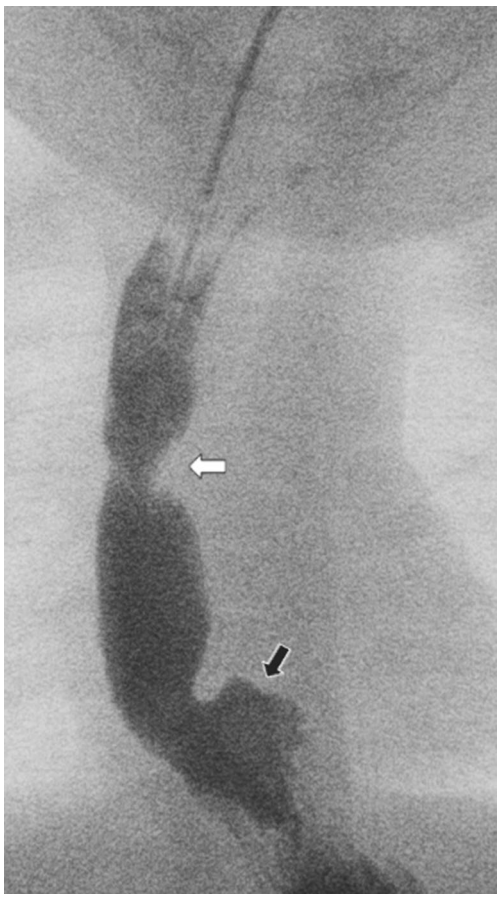

Fig. 13 Esophagram in a 4-month-old boy undergoing the Foker process 53 days after anastomosis demonstrates a hiatal hernia (*black arrow*) and narrowing at the anastomosis (*white arrow*)

Fig. 14 Esophagram after the Foker process and fundoplication for treatment of GER demonstrates an expected filling defect from gastric fundoplication (*arrow*)

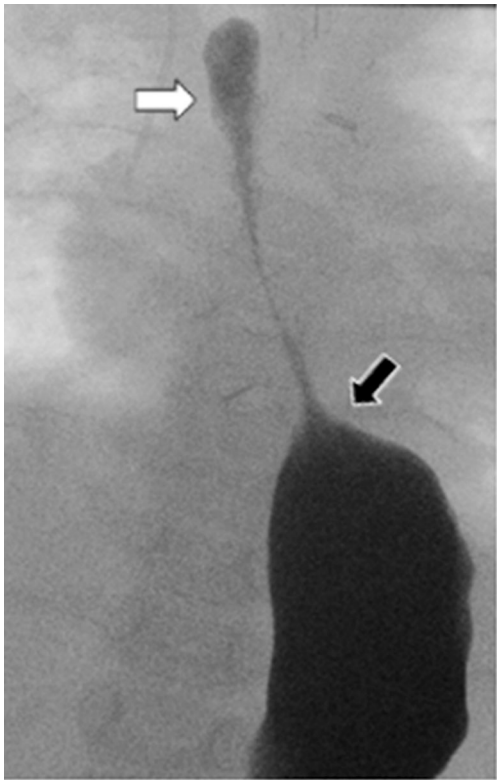

Fig. 16 Gastrostomy contrast medium injection in a 12-month-old girl postoperative day 8 after fundoplication demonstrates faint impression from fundoplication (*black arrow*) and gastroesophageal reflux (*white arrow*)

are thought to underlie this increased fracture risk. Repeat thoracotomies lead to varying degrees of chest wall deformity (Fig. 17), and should be noted because of an increased risk of scoliosis later in life.

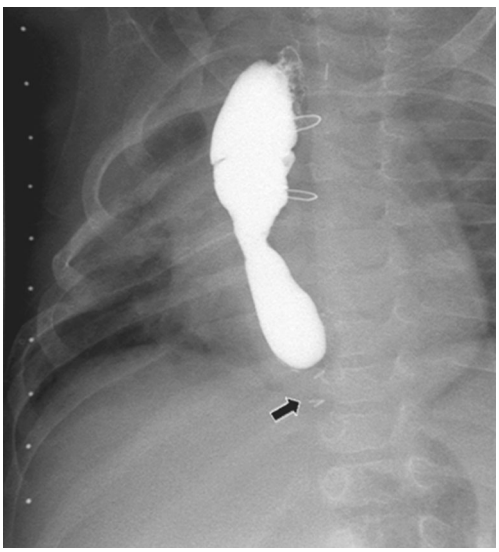

Fig. 15 Esophagram performed on postoperative day 6 after funcoplication demonstrates obstruction at the fundoplication (*arrow*) with dilatation of the esophagus and no contrast medium passage to the stomach

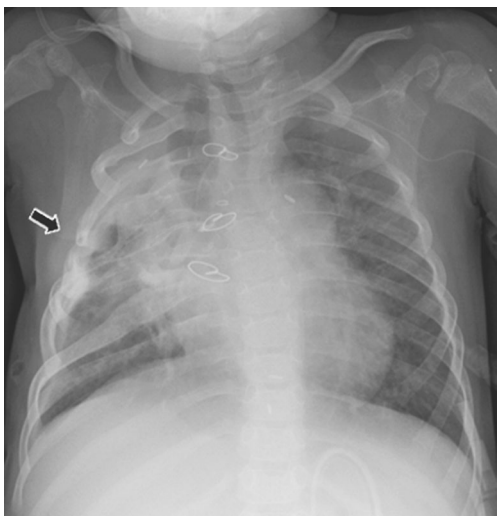

Fig. 17 Chest radiograph in a 2-year-old girl with long gap EA status post Foker process demonstrates significant rib and chest wall deformities (*arrow*) and a chronic right upper lung opacity due to plural thickening and parenchymal scar

Conclusion

Radiology is integral to the management of long gap EA utilizing the Foker process. Familiarity with expected and unexpected imaging findings in this multistage procedure will help the radiologist to provide optimum care for children.

Conflict of interest None.

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