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Pediatric Radiology

Esophageal stenosis with esophageal atresia

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Abstract. Esophageal atresia with tracheosophageal fistula may be associated rarely with distal esophageal stenosis. Three patients are reported with this combination of esophageal anomalies. In addition the clinical and radiologic features of 24 patients previously reported in the literature are reviewed. Careful evaluation of the distal esophagus during postoperative contrast studies in patients with esophageal atresia should be obtained to exclude distal stenosis. The presence of unrecognized distal esophageal stenosis may lead to complications of postoperative anastomotic leaks, poor healing of the anastomosis, aspiration, and impaction of a solid food bolus proximal to the stenosis.

Esophageal atresia and tracheoesophageal fistula (TEF) is a complex congenital anomaly. There are many types and subtypes [1]. This report presents three new patients with esophageal atresia and TEF associated with distal esophageal stenosis. In addition the clinical and radiologic features in 24 patients previously reported in the literature are reviewed.

Patient reports

Patient 1

This 13-hour-old female infant was born at another hospital where a diagnosis of esophageal atresia with distal TEF was established. The patient was referred to Henrietta Egleston Hospital for Children for further management. On the day of admission, with the patient under general anesthesia, a gastrostomy tube was inserted into the stomach. Three days later the TEF was closed surgically. Five days later a right pneumothorax developed which was treated by thoracostomy tube drainage. Cultures of pleural fluid at that time revealed the presence of E. coli. Contrast studies of the esophagus (Fig. 1a) demonstrated a small fistula between the esophagus and the thoracostomy tube. This fistulous tract closed spontaneously when the patient was treated with intravenous hyperalimentation. The contrast studies also demonstrated, for the first time, a distal esophageal stenosis, 2 cm in length, at the junction of the middle and distal thirds of the esophagus.

At 28 days of age the child underwent esophagoscopy and esophageal dilatation. The site of the anastomosis was widely patent and healing. The distal stenosis was soft and easily dilated to the size of a no. 18 French catheter. A second dilatation of the distal esophageal stenosis was performed 10 days later. The patient was discharged from the hospital at 41 days of age.

At home the child was given a diet of solid food but did not thrive well. She underwent subsequent dilatation of the distal stenosis at the age of 11 months (Fig. 1c). At 24 months of age she developed an impaction of food in the esophagus proximal to the distal stenosis. This was removed during esophagoscopy after which the stenosis was dilated. She was given only pureed foods after this episode.

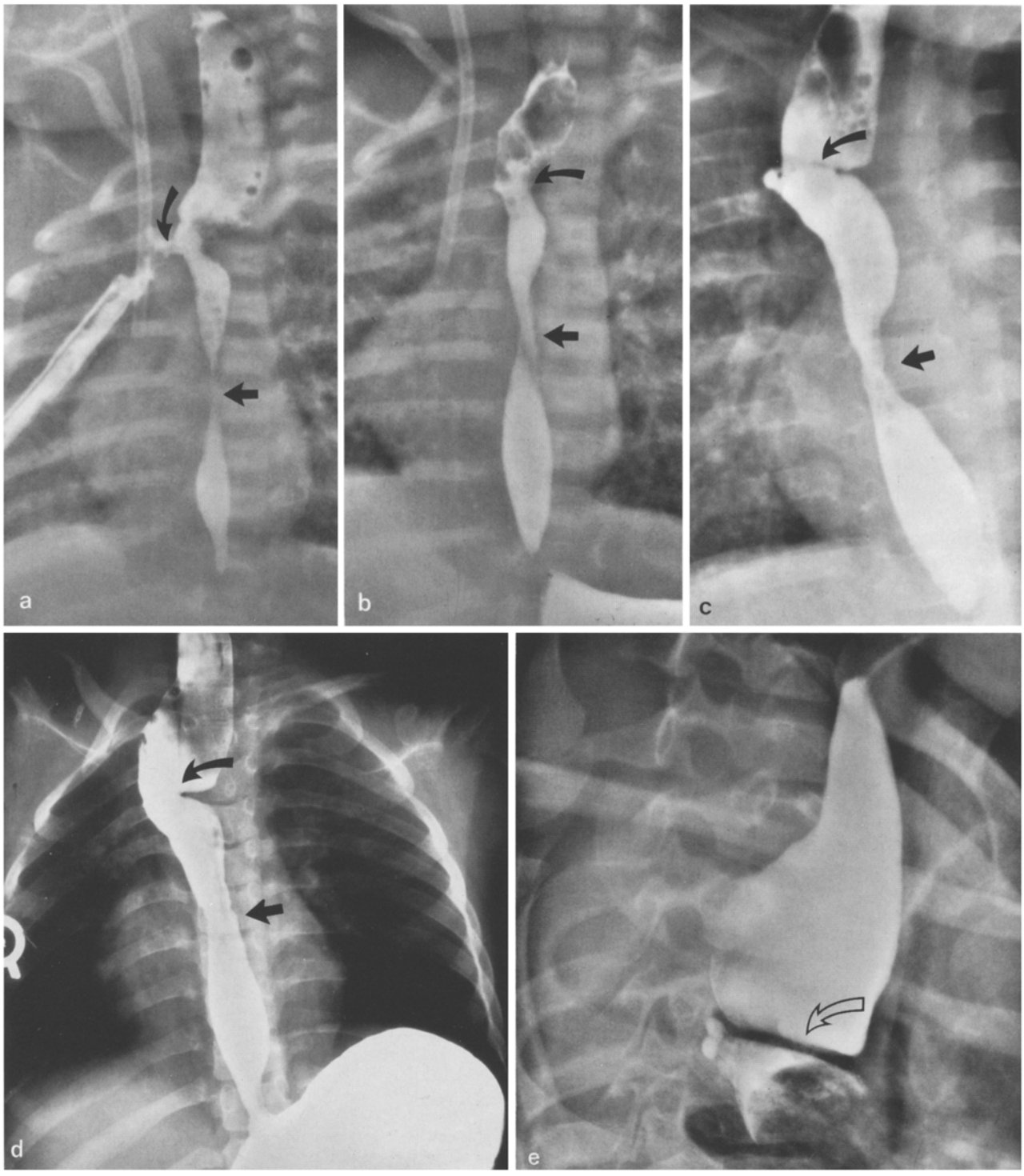
The child was admitted to the hospital at the age of 2.5 years and again at 7 years because of recurrent esophageal obstruction secondary to food impaction proximal to the distal esophageal stenosis (Fig. 1e). The esophageal stenosis was dilated on these admissions. The patient is now 9 years old with no significant clinical problems.

Patient 2

This newborn male infant was diagnosed as having esophageal atresia with distal TEF shortly after birth. He underwent a primary repair of the TEF 14 h after birth. Contrast studies of the esophagus performed 1 week after surgery revealed only slight narrowing of the anastomosis in the upper one third of the thoracic esophagus (Fig. 2). In addition an area of distal stenosis 1.5 cm in length and 3 mm in width was noted. At 2 weeks of age esophagoscopy and esophageal dilatation were performed.

Patient 3

This newborn female infant was diagnosed as having esophageal atresia with distal TEF shortly after birth. A gastrostomy tube was inserted on the day of birth and the TEF was repaired on the third day of life. A postoperative esophagram was obtained at 8 days of age. This showed slight narrowing at the anastomosis. An area of narrowing more distally was considered initially to be secondary to peristalsis. A repeat esophagram performed at 4 months of age when the child was admitted for elective repair of a right inguinal hernia revealed persistent narrowing at the junction of the middle and distal third of the esophagus (Fig. 3). The diagnosis of distal esophageal stenosis was established at this time. The proximal anastomosis in the esophagus was widely patent.



Figs. 1 a–e. Patient 1. **a** Postoperative esophagram at 3 weeks of age. A fistula (curved arrow) is present between the anastomotic site and the extrapleural thoracostomy tube. Note distal esophageal stenosis (straight arrow) at the junction of middle and distal thirds of the esophagus. **b** Postoperative esophagram at 1 month of age. Anastomosis is patent (curved arrow). The previous esophageal fistula has healed. Persistent stenosis (straight arrow) is present in distal esophagus. **c** Esophagram at 11 months of age. Anastomotic site (curved arrow) is widely patent. Persisting stenosis (straight arrow) in distal esophagus. **d** Esophagram at 5½ years. Anastomosis is patent (curved arrow). Slight residual distal esophageal stenosis (straight arrow) after dilatation. **e** Esophagram at 6 11/12 years. Anastomosis (curved open arrow) is just proximal to large filling defect due to impacted food bolus above distal stenosis

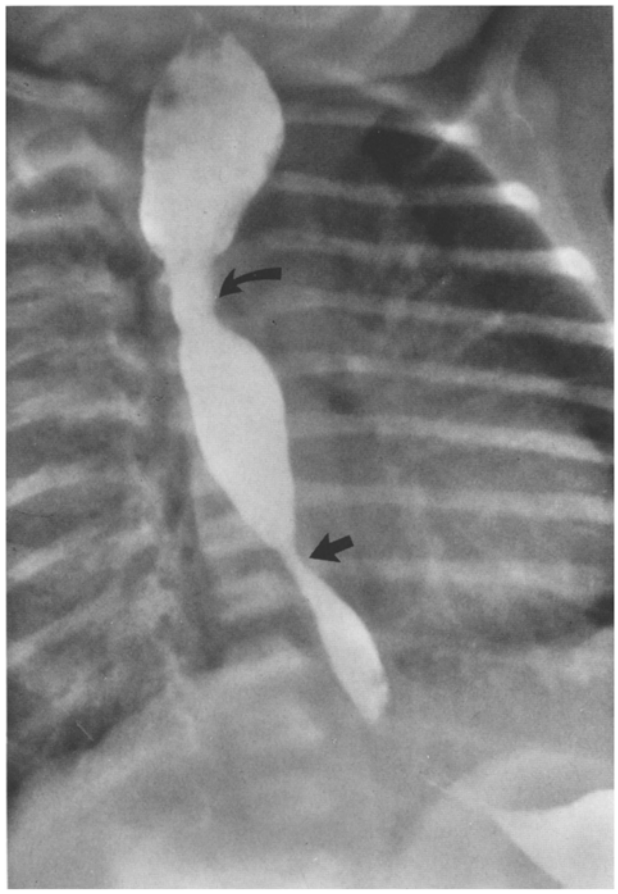


Fig. 2. Patient 2. Postoperative esophagram at 1 week of age. A patent anastomosis is present (curved arrow). A prominent stenosis is present in distal third of esophagus (straight arrow)

Review of the literature

Twenty-four patients with esophageal atresia and/or TEF in association with distal esophageal stenosis have been reported in the English literature [2–13]. In 11 of the patients the reports were detailed [2–8]; in 4 no detailed information was given [12, 13]. Detailed radiologic information was reported in 16 patients [2–7, 10, 11], clinical information in 13 [2–9], and the type of esophageal atresia in 21 [2–12]. Pathological findings were available in only 5 patients [2, 5, 6, 8].

The radiologic diagnosis of esophageal atresia/TEF with diatal esophageal stenosis is not established prior to surgery in most patients. Only in 1 patient was the distal stenosis diagnosed preoperatively [5]. The diagnosis can be suspected during surgery for esophageal atresia if a no. 8 French catheter cannot be passed through the distal esophagus into the stomach [3, 5].

Pathological changes

Histological changes in the distal esophageal stenosis have been reported in only five patients. In 3 patients the histological findings are identical. Both stenotic segments contained cartilage as well as abundant tracheobronchial seromucous glands [2, 6]. In the fourth patient the excised distal stenotic segment of the esophagus showed no histologic abnormalities [5]. In the fifth patient autopsy at 5 days of age revealed an annular stricture 2 cm in length distal to a patent esophageal anastomosis [8]. Histologically the narrowed segment consisted of a ring of abundant dense fibrous tissue.

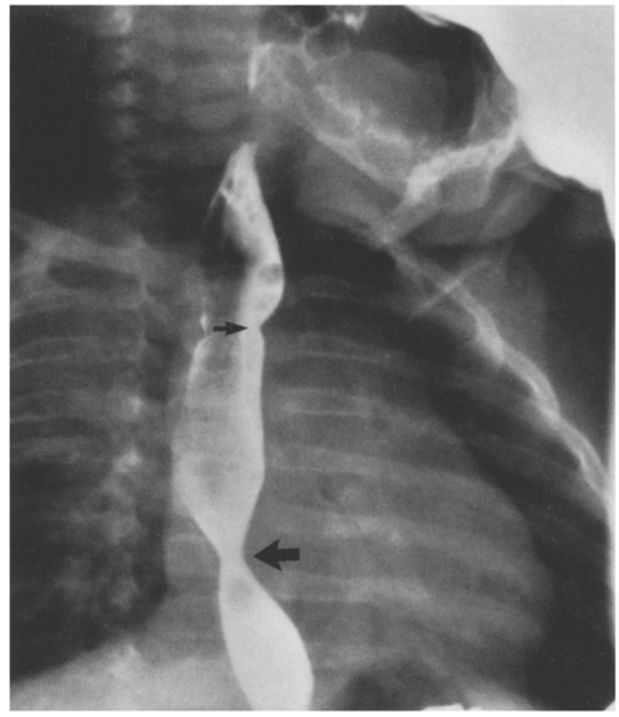


Fig. 3. Patient 3. Postoperative esophagram at 4 months of age. Widely patent anastomosis (small arrow). Stenosis in distal esophagus (large arrow)

Surgical findings

The distal esophageal stenosis was usually not seen during surgical repair of esophageal atresia and/or TEF. In only 3 patients was there specific mention that a no. 8 French catheter could not be passed through the distal esophagus at the operating table [3, 5, 8].

Radiologic findings

The diagnosis of distal esophageal stenosis in association with esophageal atresia and/or TEF was most often established by postoperative contrast studies of the esophagus (Figs. 1a, 2, 3). A persistent narrowing of the esophagus distal to the anastomosis was observed during fluoroscopy. The narrowing was interpreted initially as spasm of the esophagus in 1 patient [3]; however, on repeat examination persistent narrowing of the esophageal lumen was recognized.

Postoperative complications of esophageal atresia with distal esophageal stenosis

The postoperative course of patients with distal esophageal stenosis associated with esophageal atresia and/or TEF may be more protracted because of complications secondary to the persistent distal partial esophageal obstruction. Repeated respiratory problems were present in nine patients [2–4, 8, 9]. Anastomotic leaks

occurred in 3 patients [3, 6]. Repeated episodes of foreign body impaction in the distal esophagus occurred in 5 patients [2–4]. In 3 patients there were no respiratory or digestive problems related to the distal esophageal stenosis until the patient began to take solid food.

Discussion

Since proximal esophageal atresia with distal tracheoesophageal fistula is the most common esophageal anomaly [14], it is understandable that 17 of the 21 patients in the literature where the type of esophageal atresia was stated had the distal esophageal stenosis associated with esophageal atresia and distal TEF [2–5, 8–10, 12]. The distal stenosis was associated with isolated esophageal atresia in 2 patients [3, 6] and H-type fistula with no esophageal atresia in 2 patients [7, 11].

Isolated congenital esophageal stenosis is an uncommon cause of esophageal obstruction. There are two types of this anomaly: a web or membranous type and a segmental type [10, 15]. The segmental type of congenital esophageal stenosis may be due to fibromuscular thickening of the esophageal wall or may be secondary to tracheobronchial remnants within the esophageal wall.

Those patients with the fibromuscular thickening of the esophageal wall have a lesion which is classically one to 4 cm in length, of hour-glass configuration with smooth walls, and located at the junction of middle and lower thirds of the esophagus [15]. This lesion usually results in only partial obstruction of the lumen unless there is a secondary foreign body impaction. These stenoses can often be dilated without difficulty [16]. Although the stenosis secondary to tracheobronchial remnants may appear similarly radiologically, this lesion is usually more distal (within 3 cm of the cardia of the stomach) and more often shows high grade obstruction [17]. This type of stenosis may simulate achalasia [18] and is frequently unyielding to attempted dilatation [2, 18]. Tracheobronchial remnants are found histologically when the stenotic area is resected or biopsed. Hokama et al. [19] have recently demonstrated in autopsy material a high incidence of tracheobronchial remnants in the lower esophageal segment in patients who have no distal stenosis associated with esophageal atresia and TEF.

In all of the patients with esophageal atresia and/or TEF, including our patients, the distal esophageal stenosis was segmental. Most of these lesions were probably secondary to fibromuscular thickening of the wall of the esophagus when one considers location as well as clinical and radiologic characteristics.

Distal esophageal stenosis in a patient with esophageal atresia may be the cause of postoperative complications when the atresia is repaired. Increased pressure within the proximal esophagus secondary to the distal relative obstruction may be a factor in the development of an anastomotic perforation in the postoperative period. The chance of aspiration is also increased. Later when the patient is on a more solid diet, impaction of an ingested food bolus just above the distal stenosis is not uncommon. Dysphagia often does not occur until the child has begun to take semisolid or solid food.

The association of distal esophageal stenosis with esophageal atresia and/or TEF is uncommon. Holder [13] reported only 3 patients having esophageal stenosis associated with esophageal atresia and/or TEF in a collected series of 1058 patients with esophageal atresia variants. In a 10-year period only 3 patients with esophageal atresia and distal stenosis were seen at the Children's Hospital of Los Angeles [3]. Perreault et al. reported 3 patients out of 125 with esophageal atresia who had an associated distal stenosis [9].

When contrast studies are performed on patients after surgical repair of esophageal atresia, careful observation of the distal esophagus during fluoroscopy is essential to exclude an associated stenosis. When an associated distal esophageal stenosis is present there may be increased morbidity. Early recognition of this lesion will require more careful postoperative evaluation and early dilatation if clinically indicated.

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Literature in pediatric radiology (continued from p. 188)

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