

## Diagnostic difficulties in esophageal atresia with proximal and distal tracheoesophageal fistula: a case report

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**SUMMARY:** Ezer SS, Oğuzkurt P, Temiz A, İnce E, Hiçsönmez A. Diagnostic difficulties in esophageal atresia with proximal and distal tracheoesophageal fistula: a case report. Turk J Pediatr. 2010; 52: 104-107.

The authors report two patients with esophageal atresia and double fistula, one of whom was diagnosed intraoperatively and the other preoperatively through upper pouch contrast study. Patients with proximal and distal fistula do not present with excessive oral secretion and drooling because they can swallow the saliva/meals that reach the trachea from the proximal esophagus via the proximal fistula and then from the trachea to the distal esophagus via the distal fistula. Thus, since this clinical entity can be easily overlooked, some radiological and clinical signs that are helpful for preoperative and intraoperative diagnosis of a proximal fistula are discussed.

**Key words:** esophageal atresia, double tracheoesophageal fistula, proximal and distal tracheoesophageal fistula, recurrent pneumonia.

Of the various anatomic types of esophageal atresia, atresia with proximal and distal tracheoesophageal fistula is the least common, with an incidence of approximately 1%<sup>1</sup>. Two reports, however, have been published with much higher relative incidence of esophageal atresia with a double fistula. Dudgeon and associates<sup>2</sup> reported an incidence of 5.3% in a series of 170 patients, and Johnson and associates<sup>3</sup> reported an incidence of 7.7% in a series of 78 patients. Therefore, it would seem that the incidence of this anomaly would be much higher than previously reported if surgeons specifically looked for it. However, it can be easily overlooked.

A review of the literature demonstrates that the percentage of postoperatively diagnosed esophageal atresia with proximal fistula is higher than that diagnosed either preoperatively or intraoperatively<sup>2-6</sup>. Overlooking a proximal fistula can lead to increased morbidity and mortality owing to severe respiratory infection and the necessity for a second operation. It seems wise, therefore, to exclude a proximal fistula in all patients with esophageal atresia. Here, we present two newborns with esophageal atresia and a double fistula and indicate some findings that demand specific attention.

### Case Reports

#### Case 1

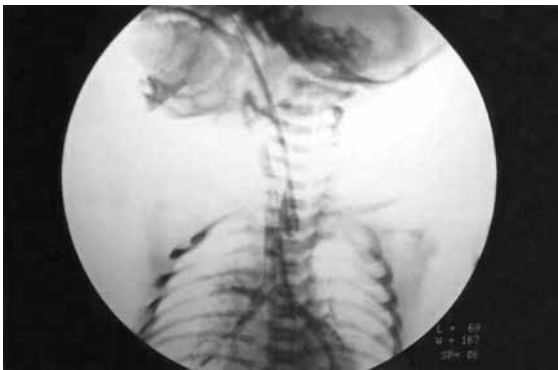
A 22-day-old female infant was referred to our hospital because of excessive salivation, inability to swallow, coughing, choking, and episodes of cyanosis that occurred since birth whenever food was offered. She was born at 38 weeks' gestation with a birth weight of 3300 g. Breath sounds were diminished in the right upper hemithorax. A nasogastric tube was blocked at 10 cm. The diagnosis of esophageal atresia was confirmed by a contrast study of the upper pouch. The results of the contrast study showed a narrow upper pouch; the tracheobronchial tree, which was interpreted as aspirated contrast medium, was also visualized.

During the operation, distention of the upper pouch was seen with each manual respiration, and after opening the upper pouch, a significant air leak was detected with a sudden collapse of the lung. A proximal tracheoesophageal fistula was detected cephalad to the end of the proximal pouch. The patient underwent an extrapleural proximal and distal fistula repair and a primary esophageal anastomosis.

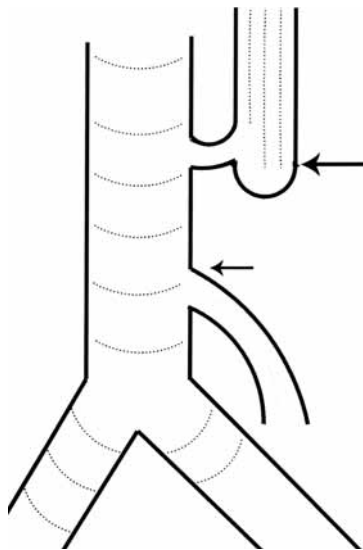
Feeding was begun through a nasogastric tube on the third postoperative day and by mouth on subsequent days because the upper gastrointestinal contrast study showed no contrast leak at the site of the anastomosis.

### Case 2

A seven-hour-old female infant with Down syndrome was admitted to the pediatric surgery department because of excessive salivation and an inability to swallow. She was born at 38 weeks' gestation with a birth weight of 3500 g. A feeding tube was blocked at about 10 cm from the nose. An upper pouch radiogram showed a narrow proximal pouch and leakage of the contrast to the tracheobronchial tree via a proximal fistula (Fig. 1a, 1b). Preoperative echocardiography showed a small ventricular septal defect and valvular pulmonary stenosis.



**Fig. 1a.** Upper pouch radiogram showing esophageal atresia and proximal tracheoesophageal fistula; air in the gastrointestinal system confirms a distal fistula.



**Fig. 1b.** The schematic picture shows esophageal atresia and proximal (long arrow) and distal (short arrow) tracheoesophageal fistula.

Proximal and distal tracheoesophageal fistula repair and primary esophageal anastomosis were performed through an extrapleural approach via a right posterolateral thoracotomy. The postoperative course was uneventful. A contrast esophagography showed no leak.

### Discussion

The exact incidence of a proximal fistula is probably underestimated because some cases initially go unrecognized, and others are reported as recurrent tracheoesophageal fistula after repair of esophageal atresia with distal tracheoesophageal fistula<sup>1-3</sup>. The lower incidence of esophageal atresia with a proximal fistula with or without a distal fistula in the literature is the result of a missed diagnosis. The pre-, intra-, and postoperative diagnosis is difficult, and more than half of the patients can be missed initially<sup>2,4</sup>. A recent review reported that esophageal atresia with a proximal and distal tracheoesophageal fistula is diagnosed preoperatively in 21.7%, intraoperatively in 34.8%, and postoperatively in 39.1% of all cases<sup>4</sup>.

Presence of a proximal fistula should be ruled out preoperatively. Preoperative diagnosis can be done by means of a contrast study of the upper esophagus, or by means of a tracheoscopy; however, even then, a proximal fistula may be missed. A contrast examination is the diagnostic method of choice to diagnose this rare proximal fistula. In the presented patients, the tracheobronchial tree was visualized during the upper pouchogram. In Case 1, it was interpreted as aspiration of the contrast medium but was proved to be due to a proximal fistula during surgery. In Case 2, the proximal fistula was clearly visualized in the pouchogram. Therefore, we suggest that a proximal pouchogram is essential for preoperative evaluation to detect a proximal fistula. Although placement of contrast medium may increase the risk of aspiration, overlooking this anomaly far exceeds the risks that come from performing a contrast study.

Gassner and associates<sup>7</sup> mention that radiographs that show contrast medium in the trachea, which is interpreted as aspirated contrast medium, should be carefully re-examined for a possible proximal fistula, as was the case in one of our patients in whom the

proximal fistula was missed during the upper pouchogram. While a dilated upper pouch is seen in patients with a distal tracheoesophageal fistula, in our patients, we showed a small, narrow upper pouch and contrast in the trachea in the presence of a proximal fistula.

Recently, ultrasound evaluation of esophageal atresia and a tracheoesophageal fistula has been advocated for defining the exact disorder<sup>7</sup>. It is postulated that mediastinal ultrasound allows documentation of the length, configuration, and structure of the wall of the blind upper pouch and the presence of a fistula by demonstrating tiny air bubbles that move in the soft tissue between the esophagus and the trachea, or ascend within the esophagus<sup>7</sup>. The disadvantage is that the technique is operator-dependent.

The symptoms, diagnosis, and treatment of an unrecognized proximal fistula are similar to those of an isolated tracheoesophageal fistula. Asthma and persistent cough over several years were the primary complaints in two patients with a history of esophageal atresia and distal tracheoesophageal fistula repair as a neonate<sup>8,9</sup>. Human saliva is composed mostly of water, but also includes electrolytes, mucus, antibacterial compounds, and various enzymes. We suggest that the first presented patient in whom the diagnosis was delayed survived for 22 days without dehydration because of passage of saliva through the proximal fistula to the trachea and through the distal fistula to the distal esophagus. However, it is still surprising that she could survive even with recurrent coughing and aspiration. Delayed recognition of a proximal fistula has been reported in the literature<sup>8,9</sup>. Islam and associates<sup>8</sup> diagnosed a missed proximal fistula using three-dimensional computed tomography in a 16-year-old girl. Because of concern about the amount of radiation being delivered to patients, the technique is currently suitable for missed cases of proximal fistula and recurrent or H-type fistulas in older patients<sup>8</sup>. Tamay and associates<sup>9</sup> diagnosed a missed proximal fistula through radionuclide gastroesophageal scintigraphy and barium swallow in a 14-year-old girl who suffered recurrent bouts of lower airway infections and chronic cough, with the diagnosis of asthma in later childhood.

Difficult mobilization of the proximal pouch may be a sign of the existence of a proximal

fistula. In fact, to localize and repair a suspected proximal fistula, the upper pouch must be mobilized completely. During the operation, the existence of a narrow upper pouch and distention of the upper pouch with each manual respiration showing continued air filling the upper pouch may be a warning sign of the presence of a proximal fistula, as was the case in one of our patients.

In conclusion, we recommend an upper pouch contrast study in all patients with esophageal atresia. We also suggest that a high index of suspicion is essential to diagnose esophageal atresia and double fistula when a newborn presents with vomiting, cough and cyanosis when offered to feed and/or inability to swallow and recurrent aspiration pneumonia with or without drooling. Those clinical findings in addition to radiological and intraoperative findings can be warning signs of a proximal fistula. These signs include a narrow upper pouch in a contrast study; leakage of contrast medium into the trachea without aspiration; distention of the upper pouch with each manual respiration during the operation; and air leak after opening the upper pouch after repairing the distal fistula. Upon presence of one or more of these findings, a search for a proximal tracheoesophageal fistula should be undertaken.

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