

Tracheoesophageal Atresia and Fistula: A Case of Mistaken Identity?

Hind Zaidan, MBBS* Ahmed Majdi Khalifa, MBCh** Isa Y Ali Hasan, MBBS, FAAP, FRCP (I), MSc***
Martin Corbally, FRCSEd, FRCS, MRCPI (Assoc)****

Tracheoesophageal atresia with or without fistula presents shortly after birth with difficulty in swallowing saliva, history of polyhydramnios and failure to pass a nasogastric tube which coils in the proximal blind ending esophagus. Early thoracotomy and primary anastomosis within a few hours of birth have produced a significant survival result; mortality is associated with coexisting serious cardiac disease, late diagnosis and presentation. The majority of patients have a fistula which could result in significant pulmonary contamination with saliva (or food) if the abnormal connection is not urgently ligated.

We report a case of a neonate referred at three days of age where the nasogastric tube (NGT) had not coiled proximally as expected; the tube initially extended vertically to the level of the diaphragm, giving the impression of an intact esophagus. The neonate subsequently underwent primary repair of the atresia with fistula ligation. At five-months post-discharge follow-up, the patient was thriving and well.

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Esophageal atresia (OA) with tracheoesophageal fistula (TEF) is a major congenital anomaly that occurs in approximately every 3,500 live births¹. It is classified according to the anatomical location and the presence or absence of the fistula between the trachea and oesophagus². Early diagnosis of an atresia and fistula is crucial in the management and prognosis and is aided by coiling of a nasogastric tube (NGT) in the proximal pouch. The presence of gas in the stomach and intestine indicates a fistula from the distal esophagus to the airway. Infants with OA are usually symptomatic immediately after birth and present with excessive secretions that cause drooling, respiratory distress and feeding difficulty.

Early diagnosis, prompt ligation of the fistula and primary anastomosis are essential to the management and have resulted in more than 90% survival rate³. Survival is determined by the presence or absence of serious associated cardiac lesions; morbidity is often determined by the presence or absence of pneumonitis secondary to late presentation and aspiration of saliva or food from the esophagus through the fistula and into the lung. Developing countries have higher morbidities, not only due to delayed presentation and management, but also due to poor resources in intensive neonatal care^{4,5}.

The aim of this case is to report an unusual presentation of tracheoesophageal atresia with fistula.

THE CASE

A case of a full-term neonate showed signs of respiratory distress and excessive oral secretions shortly after birth. The patient was saturating well on room air. A nasogastric tube (NGT) was inserted, which passed easily. A chest X-ray revealed that the NGT had reached the level of the diaphragm; therefore, it was reasonably interpreted as an intact esophagus, excluding an atresia, see figure 1. However, there was a high suspicion that the NGT was not in the right place as the course of the tube was unusual; thus, a confirmation X-ray was performed after reinsertion of another NGT, see figure 2.

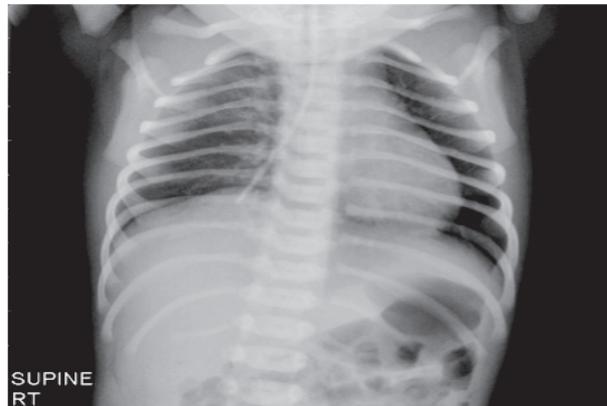


Figure 1: Unusual Placement of Nasogastric Tube

* Senior House Officer
Department of Pediatric Surgery
** Senior House Officer
Department of Pediatrics
*** Consultant Pediatrics and Neonatology
**** Chief of Medical Staff
Consultant Pediatric Surgeon
Professor and Chairman Department of Surgery, RCSI-MUB
King Hamad University Hospital
Kingdom of Bahrain
Email: hind.zaidan@khuh.org.bh

The X-ray revealed a typical image of a coiled NGT in a proximal esophageal pouch, see figure 2. A diagnosis of tracheoesophageal fistula (TEF) was confirmed, and the patient was referred for thoracotomy, ligation of the fistula and primary repair if possible. However, considering the diagnostic confusion and the convincing initial chest X-ray, the operation was preceded by esophagoscopy, which confirmed the diagnosis.

The patient was operated on the third day of life using an extrapleural approach which revealed an esophageal atresia with the proximal pouch lying high near the thoracic inlet and a distal fistula above the carina (Type C). The fistula was ligated, and primary repair was performed under some tension with the passage of a trans-anastomotic tube to allow early feeding. Despite low-pressure ventilation, the patient developed a pneumothorax 24 hours after the procedure, which required insertion of a second chest drain. A contrast swallow at five days revealed an intact repair with no leak and the patient was discharged on day 13 postoperatively on full feeds.



Figure 2: Coiled NG in Proximal Esophageal Pouch

DISCUSSION

The types of Tracheoesophageal Fistula are as follows: Type A is a pure esophageal atresia in which the proximal and distal esophageal pouches are blind, the gap is usually intermediate or long which makes primary repair difficult (7%), Type B is an esophageal atresia with a proximal fistula which varies in size from small to large. If not detected preoperatively by bronchoscopy, it could be identified intra-operatively while exploring the proximal esophageal pouch, Type C is the most common distal tracheoesophageal fistula, typically diagnosed on abdominal X-ray by the presence of gas in the stomach (85%). Type D is a proximal and distal fistula and is the rarest type (less than 1%). Type E (also called H-type) is a tracheoesophageal fistula without an atresia. These patients are usually diagnosed late, presenting with recurrent aspiration pneumonia (4%). Types A to D typically present with symptoms of choking at feeds, cough and/or aspiration pneumonia with any or none of the VACTERL associations, see figures 3 A to E.

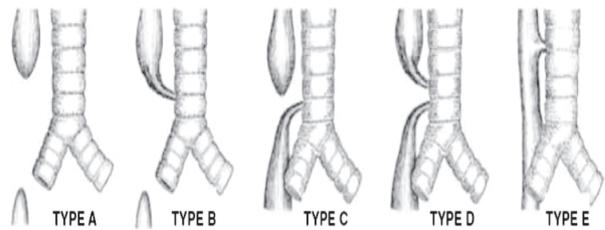


Figure 3:

Type A: Pure Esophageal Atresia

Type B: Esophageal Atresia with a Proximal Tracheoesophageal Fistula

Type C: Esophageal Atresia with a Distal Tracheoesophageal Fistula

Type D: Esophageal Atresia with a Proximal and Distal Tracheoesophageal Fistula

Type E: 'H' Type Tracheoesophageal Fistula

The presentation depends on the type of TEF and the presence or absence of a fistula, see Figures 1 A to E. Equally important are the associated conditions related to the VACTERL complex with cardiac defects being the most common (50% of cases)⁶. Antenatal diagnosis is successful in two-thirds of pregnancies and is associated with polyhydramnios. However, many cases are not detected, and diagnosis is made shortly after birth, as in our case. Clinical symptoms range from acute respiratory distress and pneumonitis to late diagnosis due to recurrent chest infections.

Pini Prato et al found that the median age of diagnosis of an H-type fistula was four days. Contrast studies were required in 4 of 5 patients, while CT chest was required in one to confirm the diagnosis⁷. A standard chest X-ray after birth, in most cases, is sufficient for diagnosing an esophageal atresia with the typical image of a coiled NGT in the proximal pouch. In some cases, the diagnosis could be challenging, and a barium contrast study provides a detailed anatomy and delineates the presence of a fistula. This is recognized as a diagnostic modality, especially in H-type fistulas. However, it must be used in a very controlled manner as to avoid the consequences of aspiration. Bronchoscopy is often used to confirm a fistula and aid in surgical repair⁸.

Delayed diagnosis impacts survival and carries prognostic implications. Waterston described pneumonia as a recognized prognostic factor as well as congenital anomalies and birth weight⁹. In developing countries, pneumonitis is a main determinant of survival, especially in centers that do not have access to skilled neonatal surgery, adequate ventilator equipment and aspiration¹⁰. Nowadays, Waterston's classification has been replaced by the Spitz classification due to improved neonatal respiratory care and ventilatory support. This relies on birth-weight and presence or absence of a major cardiac anomaly¹¹. Accurate diagnosis is imperative in early management and survival.

CONCLUSION

Misdiagnosis of a tracheoesophageal fistula results in an increased morbidity and decreased survival. Diagnosing TEF is challenging at times, especially when the patient displays vague symptoms or does not require ventilatory support; a high index of suspicion is warranted. Supplementary diagnostic tools, such as barium swallow and/or endoscopy are helpful in diagnosis and should be used appropriately in selected cases.

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