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Management of a double H-type tracheoesophageal fistula without esophageal atresia: a case report

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Abstract

Background Double congenital tracheoesophageal fistula (TEF) without esophageal atresia, known as double H-type fistula, is an extremely uncommon malformation. Due to its nonspecific symptoms and the small size of the fistula in infancy, H-type tracheoesophageal fistula is rarely diagnosed in the neonatal period and is also challenging to repair.

Case presentation We report a rare case of double H-type tracheoesophageal fistula in a 2-day-old newborn, identified with a time-lapse and repaired separately. The diagnosis was established through repeated bronchoscopy, with methylene blue injected through an esophagoscope. A right thoracotomy was performed to undertake the surgical repair after cannulating the fistula with a vascular guide wire. Five months later, the infant, who had a history of recurrent episodes of coughing and shortness of breath, was diagnosed with acute respiratory distress, and a second TEF was detected by bronchoscopy. A cervical incision was used to repair the second H-type fistula.

Conclusions Double H-type tracheoesophageal fistula should be considered while correcting a tracheoesophageal fistula, especially if the symptoms continue after the first operation. Repeated radiological contrast studies and bronchoscopy with methylene blue injected through an esophagoscope help the early diagnosis.

Keywords Double H-type tracheoesophageal fistula, Bronchoscopy, Newborn, Case report

Background

Isolated tracheoesophageal fistula, or H-type tracheoesophageal fistula, (TEF) is a rare congenital disease affecting 4 to 5% of the general population of newborns worldwide [1, 2]. An early diagnosis is still a challenge for pediatricians because of the non-specificity of symptoms [3, 4]. TEFs are frequently attributed to more common etiologies, like gastroesophageal reflux (GER), swallowing disorders, or poor feeding techniques. The double H-type fistula is even less common, and very few numbers of patients have been reported in the literature [5]. Diagnosis and management of this rare condition are real challenges. We describe a neonatal case of double H-type fistula that has been identified with a time-lapse and operated on separately, and we propose a diagnostic approach to prevent unnecessary delays in the management of this condition and ensure a better prognosis.

Case presentation

A term female infant, born at 39 weeks 2 days to a 31-year-old GIPI mother by normal spontaneous vaginal delivery after an uncomplicated pregnancy, was admitted

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to the neonate intensive care unit for respiratory distress occurring at 36 h after birth. She presented cyanotic episodes and coughing with feeds. The maternal prenatal screening results were negative. There was no history of consanguinity in the family.

On admission, the patient had respiratory distress signs with polypnea, nasal flaring, and intercostal retractions. Clinical examination revealed abdominal distension. There were no episodes of bilious vomiting, intestinal obstruction signs, or fever. A thoracoabdominal x-ray revealed aspiration pneumonia with gastric dilation. The nasogastric tube was successfully implanted. She required conventional mechanical ventilation for 24 h with good evolution. We performed an upper gastrointestinal contrast series at the age of 6 days. The diagnosis of gastroesophageal reflux disease was ruled out, with no evidence of a tracheoesophageal fistula. A second upper gastrointestinal series was repeated at the age of 11 days. It showed opacification of the tracheobronchial tree, suggesting a connection between the airway and digestive tract (Fig. 1).

With high suspicion of H-type TEF, we performed bronchoscopy with methylene blue (MB) injection via esophagoscopy. As expected, bronchoscopy above the carina revealed MB, showing a 5-mm type H bronchoesophageal fistula between the esophagus and trachea, 2 cm above the carina at the level of the third thoracic vertebra (Fig. 2). We then cannulated the fistula using a vascular catheter that facilitated intraoperative identification of the fistula.

On day 17 after birth, a right thoracotomy was performed through the fourth intercostal space. We identified the trachea and esophagus, then definitively identified the fistula, which was transected and closed with nonabsorbable interrupted sutures. Routine esophagography was performed on the 7th day after the operation. It showed no leaks and no evidence of proximal H-type TEF. The patient tolerated oral feedings and was discharged home 10 days later.

At 5 months of age, the infant developed severe shortness of breath. She had recurrent episodes of coughing and shortness of breath with wheezing for a month. Respiratory examination revealed diffuse polyphonic dry sounds and severe crackles bilaterally. Chest and abdomen radiographs showed aspiration pneumonia with marked gastric distension. Recurrence of TEF or another H-type fistula was suspected. An upper GI series was planned and revealed a 5-mm fistula between the upper third of the esophagus and the trachea (Fig. 3). Therefore, rigid bronchoscopy under general anesthesia was planned to identify the lower fistula forming a pouch, the second H-shaped fistula. This fistula was very high, above the second thoracic vertebra. We cannulated it with a vascular guide wire. Surgery was recommended. We performed a cervical approach. The fistula was deep in the throat, high at the entrance to the chest. It was divided near the esophagus. The trachea was closed first, then the esophageal side of the fistula was also closed. Then, we insert the sternothyroid muscle flap between the two opposing suture lines.

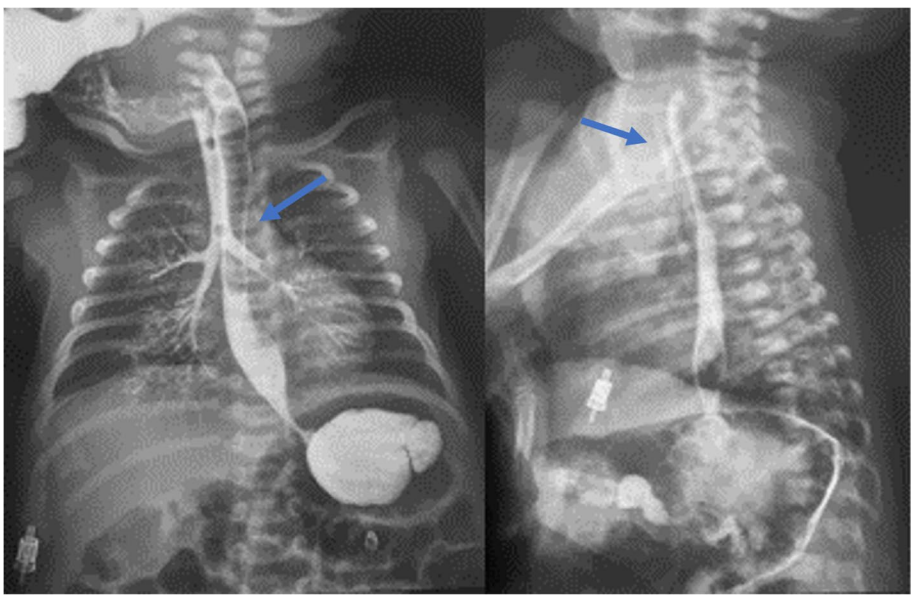

Fig. 1 Upper gastrointestinal contrast series at the age of 11 days showing the opacification of the tracheobronchial tree suggesting a connection between respiratory and alimentary tracts (blue arrows), or pulmonary aspiration

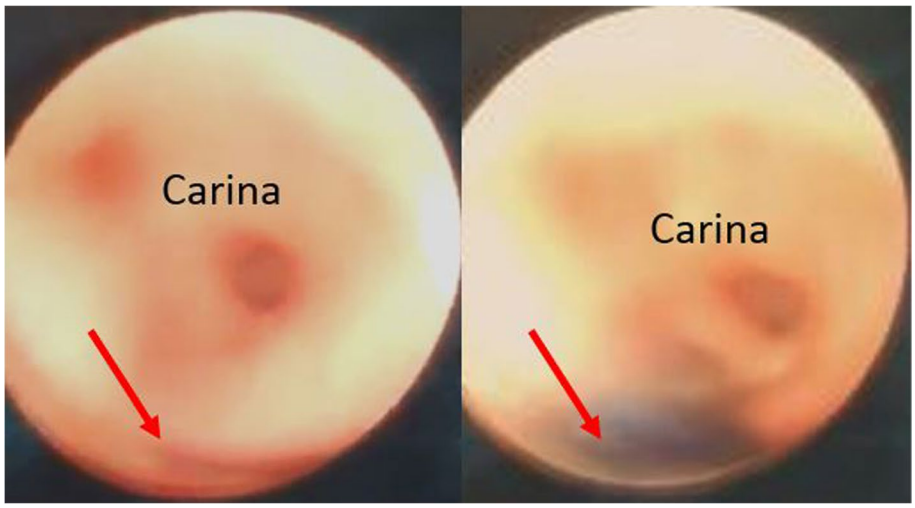

Fig. 2 Bronchoscopy showing a 5-mm H-type broncho esophageal fistula (red arrows) between the esophagus and the trachea located at 2cm above the carina after Methylene Blue instillation

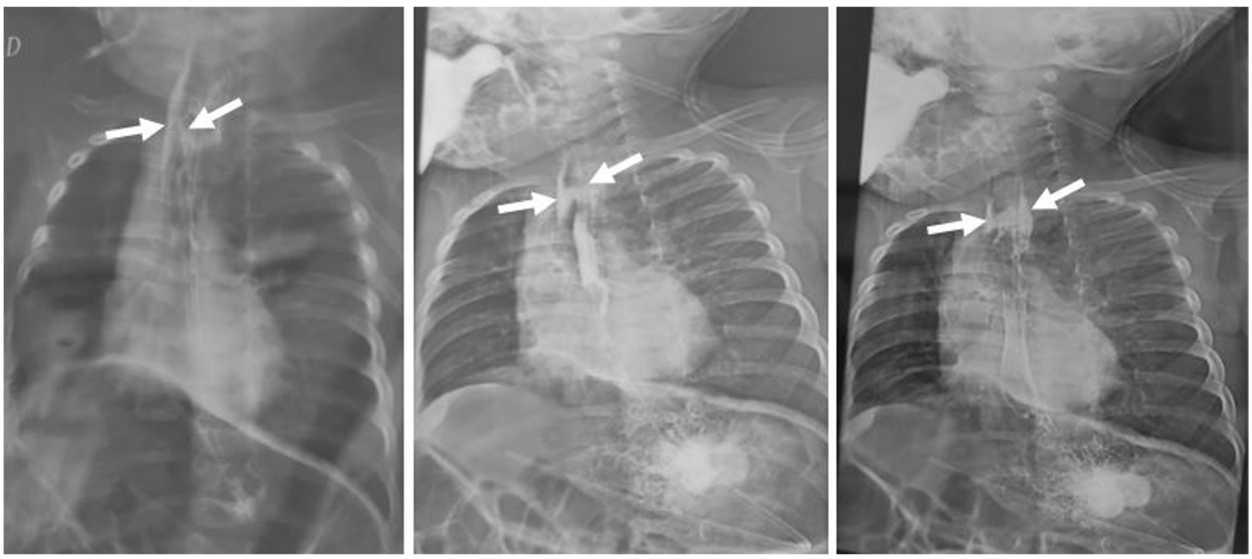

Fig. 3 Upper gastrointestinal series showing a 5-mm fistula between the upper third of the esophagus and trachea (arrows). Substantial material: video of bronchoscopy with methylene blue injection via esophagoscopy

During the postoperative period, the patient showed a good course, and an esophagogram performed on the sixth postoperative day showed no leaks. The patient was discharged on day nine after the introduction of oral nutrition.

Discussion

Congenital TEF occurs due to a defective separation of the respiratory and digestive tracts during embryogenesis, leading to abnormal communication between the two [6]. The incidence of isolated TEF or H-type TEF is very rare, approximately 1/50,000 per 100,000 live births, with a slightly higher prevalence in males [7]. The clinical diagnosis of a fistula is often quite difficult because it can appear with various symptoms of different severity. Therefore, an early diagnosis is still a challenge in the neonatal period and may be delayed into childhood or even into adulthood [1, 4].

Double TEFs without atresia of the esophagus have a very low incidence rate. There are few reports of patients having more than one fistula [8–14]. To our knowledge, this is the first case with double H-type TEF in a Tunisian

newborn who has been operated on first with a preoperative diagnosis of an isolated H-type fistula located 2 cm above the carina at the level of the third thoracic vertebra. And then, another H-type fistula was located above the level of the first lesion. Among children, this is only the sixth case to be reported in the medical literature. Table 1 summarizes the literature on pediatric double H-type TEFs.

The classical clinical triad of H-type TEF called the triad of Helmsworth et Pryles, includes coughing with cyanosis and choking on feeding, abdominal distention, and recurrent pneumonia [1, 2, 5, 13]. Typically, symptoms start from birth, but the diagnosis is easily missed in the neonatal period [2, 3]. These symptoms are not specific, and they are shared with other disorders like laryngeal cleft, GER, and swallowing disorders [1, 4]. Difficulty in diagnosing double H-type TEF in the literature might be due to extremely low incidence and difficulty in the visualization of fistulae in a single diagnostic study. To make a correct diagnosis, starting with a high index of suspicion is necessary because symptoms are almost the same as in a single H-type fistula. In our case, we found such classical symptoms. They appeared after the first surgery; therefore, the diagnosis of a recurrence or a second TEF was suspected.

Diagnostic methods in suspected H-type fistulae are upper gastrointestinal series and esophagogram proceeded with water-soluble contrast. Their sensitivity ranges from 50 to 73% [7]. CT scan is recommended to assess the pulmonary parenchyma to rule out a fistula. However, it is not the first line study to detect a congenital TEF due to radiation exposure, especially in younger patients. Esophagoscopy is less useful in TEF identification since the esophageal ostium is smaller, and the intraluminal air-pressure during the digestive endoscopy may intermittently close the opening with mucosal folds [3].

Bronchoscopy represents the most reliable tool regarding clinical TEF suspicion. It confirms the diagnosis, establishes the level of the fistula, and identifies associated tracheobronchial anomalies and any additional fistula [3, 15]. Huaying Li et al. [15] stated that the use of a bronchoscopy tracer via esophageal MB injection could be made in the neonate as it is an effective method of confirming the diagnosis and may avoid prolonged suffering. A catheter can be inserted through the fistula to facilitate its intraoperative identification [7]. In our case, the use of a bronchoscopy tracer via esophageal MB injection was able to pick up the first fistula. However, we missed the second one at the first operation. So, the examination of the trachea by bronchoscopy is recommended and must be done carefully. To improve the test's performance, it is recommended to instill MB using a catheter placed in the esophagus. The presence of a fistula is confirmed if MB passes through the fistula into the trachea.

The treatment of choice is surgical repair. The cervical approach is indicated for proximally located fistula, above the second thoracic vertebra, whereas the thoracic approach is recommended for those at a more caudal level [3, 16]. Surgery consists of ligation and division of the fistula and repair of the tracheal and esophageal walls. In most cases of reported double H-type TEFs, surgical repair was done subsequently. Mattei and al (9) reported the correction of one such fistula in one surgery. The thoracic fistula was corrected by surgical repair, and the cervical fistula was corrected by a balloon catheter. In

Table 1 Reported pediatric cases of double TEFs without esophageal atresia

Case	Year	Age	Sex	Symptoms	Pre-operative imaging	Diagnosis of the second TEF	Outcome
1	1952 (12)	10 days	F		–	Postmortem	Letal
2	1957 (13)	2 days	M		Chest X-ray Bronchoscopy	Postmortem	Letal
3	2009 (14)	20 days	M	Recurrent vomiting Coughing, the oral intake was inadequate	Chest X-ray Bronchoscopy	7th day after the first surgery	Uneventful
4	2012 (9)	N.K	M	Coughing with feeds	Esophagram rigid Bronchoscopy	Identified in 1 operation	Uneventful
5	2018 (8)	5 days	M	Aspiration pneumonitis	Upper GI series Bronchoscopy	146th day of life	Vocal cord palsy and hypopharyngeal motility disorder disappeared at the age of 1 year
6 Present case	2021	11 days	F	Cyanotic episodes and coughing with feeds, abdominal distension	Upper GI series Bronchoscopy	At 5 months of age	Uneventful

F Female, *M* Male, *N.K.* indicates not known, *GI* Gastrointestinal

our case, the double TEF was closed separately; the first was closed by a thoracotomy, and for the second TEF, we performed a cervical approach as it was located high in the thorax.

Conclusions

Double H-type fistula is an extremely rare subtype, and delayed diagnosis may lead to management problems. It should be considered while correcting a tracheoesophageal fistula. If the symptoms continue after the first operation, the diagnosis of another fistula must be ruled out. Diagnostic procedures such as repeated radiological contrast studies and bronchoscopy with methylene blue injected through an esophagoscope can improve the rate of early diagnosis.

Abbreviations

TEF Tracheoesophageal fistula
GER Gastroesophageal reflux
MB Methylene blue
CT Computed tomography

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Not applicable.

Authors' contributions

CR: conception, data analysis, draft the manuscript, revised the manuscript, and approved the submission. TC: draft the manuscript, revised the manuscript, and approved the manuscript. MBD: conception, data analysis, revised the manuscript, and approved the manuscript. MA: data analysis, revised the manuscript, and approved the manuscript. NK: acquisition and analysis, revised the manuscript, and approved the manuscript. ABT: revised the manuscript and approved the manuscript. RM: revised the manuscript and approved the manuscript. NH: revised the manuscript and approved the manuscript. The authors have read and approved the manuscript.

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Declarations

Ethics approval and consent to participate

Not applicable.

Consent for publication

The parents of the patient have consented to the use of clinical photographs for publication and research process.

Competing interests

The authors declare that they have no competing interests.

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