



11 Cases of H-Type Tracheoesophageal Fistula: How Diagnosis and Treatment May Be Improved

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Abstract

Congenital H-type tracheo-esophageal fistulas (TOF) or esophageal atresia type IV (Vogt classification 1929) are rare congenital malformations. 1:50 000- 1:100 000 children are born with H-TOF [1]. Late diagnosis is typical and usually requires intensive diagnostics. The rareness of this malformation as well as the unspecific symptoms are the main reasons for late diagnosis. During the years 2009-2022, 11 patients with H-TOF were treated in our clinic. In this paper all cases will be presented, and diagnostics and treatment will be discussed. The aim of this study is to improve diagnosis and to minimize complications, such as paralysis of the recurrent laryngeal nerve.

Keywords: H-type tracheo-esophageal fistulas; esophagogram; tracheobronchoscopy; diagnosis

Introduction

One of the most common classification systems of esophageal atresia's is the Vogt classification. The Vogt classification divides the esophageal atresia's in types I, II, IIIa, b, c and IV. Type IV is characterized by an H-type fistula (H-TOF) between the esophagus and the trachea without atresia. H-TOFs account for 4-5% of all esophageal atresia's [1,2] with male and female being equally affected. Transtracheal surgical repair of an H-TOF was first performed in 1938 on a 6-year-old girl [3,4]. In 1948, Haight reported an H-TOF repair through a thoracic approach. In 1958, Miller reported the first cervical access repair of H-TOF. One of the greatest risks when treating H-TOF surgically is injury of the right recurrent nerve causing paralysis of the vocal cords, though the paralysis is often reversible [1,5]. Refistulas are rare. H-TOF may present with choking during feeding sessions, coughing attacks or desaturation while drinking, as well as recurrent respiratory infections. The symptoms are sometimes misleading, which may lead to delayed

diagnosis. Thus, the time of diagnosis varies from infancy to adulthood [6-11]. Diagnostic measures comprise an esophagogram with a contrast agent or tracheobronchoscopy. Due to the oblique course of the fistula- from esophageal caudal to tracheal cranial imaging with a contrast medium is difficult. Several studies have shown that imaging must occasionally be performed multiple times to diagnose an H-TOF. This poses additional risks of aspirations and radiation exposure. Tracheobronchoscopy, in contrast is a highly reliable diagnostic tool that identifies H-TOF in infants and newborns. Furthermore, using tracheobronchoscopy the fistula can be marked with a wire that helps a surgeon to identify the fistula during subsequent open repair surgery [10,11].

Patients and Methods

From 2009-2022 11 patients with H-TOF were treated in the department of pediatric surgery in Olgahospital Stuttgart. Patient

variables including gender, week of pregnancy at birth, birthweight, other malformations, symptoms, success of esophagogram, age at time of surgical repair, age at tracheobronchoscopy, complications, length of stay in hospital after surgical repair are given in Tables 1 & 2. Of the 11 patients 9 were born on time. Two were delivered

prematurely in 35+1 and 27+5 gestational week respectively. Three of the patients had other malformations- one was born with a gene defect, the other had a complete laryngeal atresia. One of the patients was born with various malformations of both feet as well as an ASD. One patient had an anal atresia.

Table 1: Patient Characteristics.

case	m/f	Week of gestation	Other malformations	Symptoms
1	f	40+1	none	recurrent coughing, especially in the night
2	m	40+1	none	recurrent cyanotic episodes after drinking
3	f	39+	VUR °IV, diagnosed at an older age	Coughing attacks, dystrophia, cyanosis while drinking
4	m	38+1	complete glottis stenosis: H-TOF saved his life	-
5	m	38+5	none	noticeable breathing, recurrent choking, cyanotic episodes while drinking
6	m	36+6	none	gasping breathing while drinking, aspiration
7	f	39+6	ASD, malformations both feet, syndactylie D2/3 both feet, hip dysplasia Ib	Recurrent coughing while drinking; fever
8	w	35+1	pyloric stenosis, twin, low birthweight (1750g)	swallowing disorder, recurrent aspirations
9	f	38+3	anal atresia, thymushypertrophia, caudal regression syndrome/ tethered cord	swallowing disorder, recurrent aspirations
10	f	27+5	de novo mutation TOP2B-gene, epilepsy, twin	swallowing disorder, recurrent infections, and aspirations
11	m	unknown	none	Recurrent infections treated as "infect asthma"

Table 2: Procedures and Results.

Case	Age at time of surgery	Tracheo- bronchoscopy before operation	Fistula seen in esophago-gram	complications	Length of hospital stay after surgery in days
1	146 d	6 days before surgery	no	atelectasis right upper lobe	10
2	14 d	6 days before surgery	yes-another clinic	none	12
3	5 d	intraoperative	yes	none	Unknown *
4	81 d	6 days before surgery	not done	Pneumonia	Unknown *
5	18 d	3 days before surgery	no	paralysis of right recurrent nerve	21
6	23 d	intraoperative	not done	none	Unknown *
7	29 d	2 days before surgery	not done	pneumonia, insufficient ventilation right upper lobe	Unknown *
8	15 d	1 day before surgery	no	none	33
9	9 d	intraoperative	not done	paralysis of right recurrent nerve	16
10	59 d	intraoperative	yes	reversible paralysis of right recurrent nerve	12
11	17y 11mo	22 days before surgery	not done	none	length of hospital stays not related to H-TOF

*Patient was transferred back to their home hospitals before discharge.

Of the 11 patients, two were born in Stuttgart. All the other patients were transferred from other hospitals. One patient had been diagnosed with H-TOF at the clinic in which the patient was born. All other patients were diagnosed in domo with H-TOF. One patient was diagnosed immediately after birth as he had a complete

larynx stenosis and would not have been able to survive without H-TOF. H-TOF was life- saving in this case. In 6 of these patients a single esophagogram was performed, which diagnosed H-TOF in 3 cases. In all patients tracheobronchoscopy was performed and the fistula was identified in all cases (Figures 1-3). One patient was

17 years old when H-TOF was diagnosed. He had a long history of recurrent airway Infections, which was misdiagnosed as infectious asthma. At the age of 17 he was treated for lymphoma and suffered from a pneumomediastinum. Thus, a tracheobronchoscopy was

performed, which revealed a large H-type Fistula. In three cases surgical repair of H-TOF was performed under the same anesthesia induced for the tracheobronchoscopy.

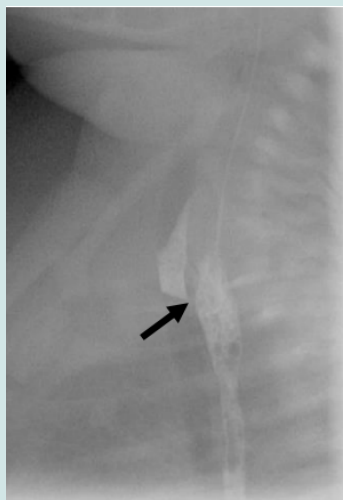


Figure 1: H-TOF in esophagogram. Contrast medium was applied to the esophagus and the contrast medium passed into the trachea through the fistula. The black arrow marks the fistula.



Figure 2: Esophagobronchial fistula (black arrow). Left bronchus (white arrow) and right bronchus (blue arrow).

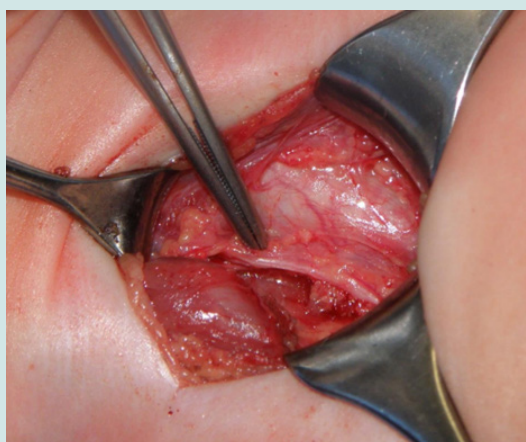


Figure 3: Right Recurrent Nerve in the Field of Surgery, marked by Forceps.

Surgical repair was usually performed within 6 days following diagnosis, with one exception- the 17-year-old cancer patient. The repair was in all cases carried out through a right cervical access. In all cases the right recurrent nerve was identified, such that maximum care could be taken to avoid damage. In infants the right recurrent nerve runs more distal to the esophagus compared with adults and therefore typically leads directly through the field of surgery, close to the esophagobronchial fistula (Figures 4 & 5). Furthermore, preparation must also be done very carefully in order not to harm the left recurrent nerve by accident. In only

one of the patients a gastroscopy had to be performed 19 months after primary surgery, when the patient presented with a stenosis. This patient underwent gastroscopy and dilatation once and has not suffered any problems related to the condition to date. Despite all caution, three patients suffered paralysis of the right recurrent nerve. One patient was lost on follow-up, the second recovered spontaneously, the third is awaiting its checkup endoscopy. Three patients suffered from pneumonia postoperatively. It is unknown whether pneumonia was a consequence of the operation or a complication of the fistula itself.

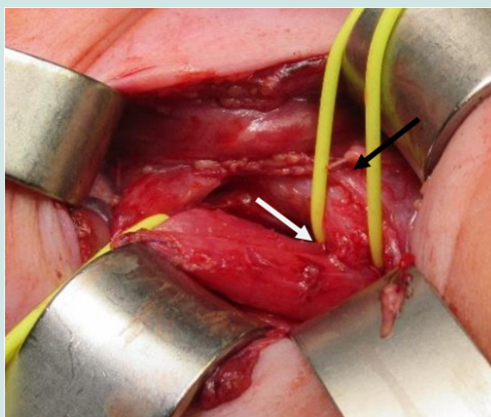


Figure 4: H-TOF (white arrow). N. larynges recurrence (black arrow) also depicted in the field of surgery.

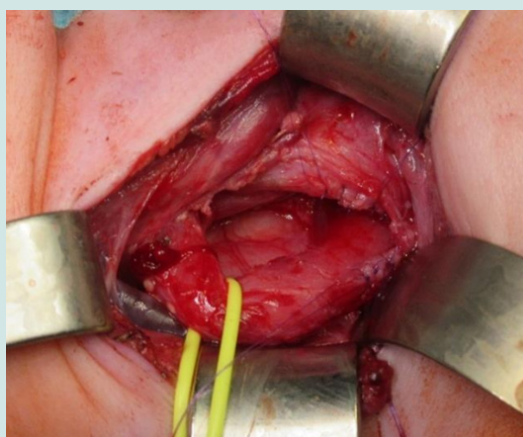


Figure 5: Field of Surgery After Dissection of H-TOF.

Discussion

Key ongoing discussions on H-TOF concern which diagnostic methods are necessary, how to diagnose H-TOF as fast as possible, what the various risks of the different methods are and how best to proceed when H-TOF is diagnosed [6,7,10,12]. In this investigation, 6 out of 11 patients had an esophagogram, which was successful in 3 cases. Not more than one esophagogram was performed on each patient. If an esophagogram was done earlier on in another clinic without success, the investigation was not repeated. The sensitivity of esophagogram was low as it was also described in many other studies before [1,6,13,14,16]. The esophagogram allows to rule out

differential diagnosis such as esophagus stenoses, gastroesophageal reflux, esophageal diverticula or swallowing disorders. These differential diagnoses can't be seen in a tracheobronchoscopy. In order not to miss differential diagnoses and at the same time not to prolong diagnosis one should perform a single esophagogram when H-TOF is suspected.

A tracheobronchoscopy was performed in all patients and was diagnostic in all. Thus, the tracheobronchoscopy is a very sensitive diagnostic tool for H-TOF. Other advantages of a tracheobronchoscopy compared to an esophagogram are a lesser risk for aspiration and no radiation. Also, there are some

rare differential diagnoses which can be identified or ruled out by tracheobronchoscopy only. Namely these are laryngeal cleft, subglottic hemangioma, or laryngeal stenosis [15, 17-21]. The disadvantage of a tracheobronchoscopy of course is, that the child needs full anesthesia. Based on the known success rate of tracheobronchoscopy for H-TOF diagnosis compared with esophagogram diagnosis observed in this study one could argue that on urgent suspicion of H- TOF, one should first resort to tracheobronchoscopy and to an esophagogram second and only if further investigation of a negative tracheobronchoscopy is required [6,10,12].

In cases of unspecific symptoms, esophagogram should still be the first diagnostic performed. In four cases surgical repair could be performed in the same anesthesia as the initial tracheobronchoscopy. In two of these patients, H-TOF was suspected as a result of a prior esophagogram. The "single-anesthesia pathway" requires a team of highly specialized ENT Doctors, pediatric surgeons, anesthesiologists, and specialized equipment. For this reason, diagnosis and repair can rarely be performed in one session only. However, increasing the proportion of patients who can be treated and diagnosed under the same anesthesia is desirable, because the fistula only needs to be depicted by the ENT doctors once, which ultimately entails less manipulation and a lower risk of injury. The therapy goal can thus be achieved faster. Studies described a vocal cord paralysis in up to 50% of operated H-TOF. In this investigation only 3 of 11 presented with a right sided vocal cord paralysis after the surgery. This comparatively favorable result may reflect the benefits of placing particular focus on preparing the right recurrent nerve when repairing for H-TOF surgery. Even if in many cases the paralysis is temporary and not permanent.

Conflict of Interest

None.

Ethical Approval

None.

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