Endoscopic Airway Evaluation in Congenital Tracheoesophageal Fistula
Pediatric airway endoscopy

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1. Abstract:

Introduction. The communication between the trachea and esophagus is called tracheoesophageal fistula (TEF). It can occur as a congenital malformation (0.025-0.05%) (in particular related to the esophageal atresia) or can occur as an acquired pathology. Endoscopic evaluation is the gold standard for the diagnosis of TEF and must be performed, in presence of symptoms such as choking, coughing, and cianosis at feeding.

Materials and methods. The authors present 145 endoscopic airway evaluations, performed in 142 children for the suspected presence of TEF and for a diagnostic classification of esophageal atresia. The endoscopic airway procedure was performed with the rigid endoscopy technique, in general anesthesia and spontaneous ventilation, with topical anesthesia.

Results. The use of the rigid endoscopy allows us to assure an open airway and assists operative management: in the presence of TEF the endoscopic procedure was in fact diagnostic, and operative at surgery. The tracheobronchoscopic airway evaluation was able to identify the presence, the level and number of TEF in all patients, in order to classify the cases and plan the therapeutic strategy. Endoscopy showed the fovea of TEF in different positions, in the upper, medium and lower part of the trachea, in rare cases a double fistula or in some cases did not detect the presence of fistula.

Discussion and Conclusions. The fovea located in the upper part of the trachea was always of small size, and difficult to diagnose, while the fovea located in the lower or medium part of
the trachea was always of large size, and simple to identify. The identification of the precise anatomic position of the TEF guides the surgical planning but also permits to achieve the optimal ventilation and strategies to reduce potential complications during anesthesia.

2. Introduction

Tracheoesophageal fistula (TEF) is an esophageal pathology that consists of the communication between the trachea and esophagus. It can occur as a congenital malformation (0.025-0.05%) classified in the esophageal atresia chapter: incomplete formation of the esophagus is the most frequent association with TEF, which may be an isolated malformation. The first patient to survive a congenital esophageal anomaly was born in 1931 with a TEF and no atresia. TEF can also occur as an acquired pathology (blunt traumatic injuries, neck or thoracic surgery, post prolonged intubation with tracheal cuffed tube associated with rigid nasogastric tube). TEF is commonly (17-70%) associated with other congenital malformations (cardiac, gastrointestinal, genitourinary, muscular and skeletal) but isolated TEF can also be present.

Endoscopic evaluation is the gold standard for the diagnosis of TEF and must be performed even if imaging techniques reveal the communication between trachea and esophagus.

In 1981 endoscopy was used for the first time in the newborn affected by esophageal atresia (EA) for the purposes of obtaining a correct classification (1). Then, in 1982, the insertion of a Fogarty catheter for the occlusion of TEF was described to facilitate the lung mechanical ventilation of an infant with EA and TEF complicated by severe RDS before the surgical closure of fistula (2).

The esophagus and trachea develop from primitive foregut and at 4-5 weeks (gestational age), the separation of the caudal part (trachea) from the dorsal part (esophagus) may be incomplete and a TEF can result.

Prenatal ultrasonography attempts cannot offer a real prenatal diagnosis: the suspicion of an esophageal atresia may arise when polyhydramnios is associated with the absence of the stomach bubble. Furthermore, in all newborns, but also in children, the presence of symptoms such as choking, coughing, and cyanosis at feeding may point to the need to evaluate the presence of a possible TEF.

The aim of this study is to evaluate the use of the airway endoscopic approach to investigate suspected symptoms of TEF, in the presence of EA at birth, before surgical repair, in recurrent fistula and to aid in performing possible endoscopic conservative closure.

3. Materials and Methods

From January 1990 to October 2013, 145 endoscopic airway evaluations were performed in 142 children for the suspected presence of TEF and for a diagnostic classification of EA (Table I). 134 patients underwent the perioperative path entirely at our center, 8 were admitted after being treated in other hospitals.
Table 1: Congenital pathology associated with TEF

Of the neonates treated entirely in our hospital, 79 (58%) were males and 55 (42%) females, 99 (74%) were born at term, 35 (26%) preterm; the mean birth weight was 2.8±0.8 Kg (range 4.1-1.1 kg). The 8 patients admitted after treatment in other centres were 5 infants (2 aged one month, 2 aged 2 months, 1 aged 3 months) and 3 children (1, 2 and 4 years old respectively).

78 patients (54.9%) had one or more other congenital anomalies (Table 1). The highest percentage consisted of congenital heart diseases (Table 2). In addition 7 cases were classifiable in a WACTERL syndrome, while 2 in a CHARGE association.

The endoscopic airway procedure was performed with the rigid endoscopy technique, in general anesthesia and spontaneous ventilation, with topical anesthesia of the pharynx, larynx and trachea (lidocaine 4-5 mg/kg).

In neonatal age, the rigid ventilating bronchoscope (Storz® 2.5 or 3 mm Internal Diameter, 180 mm length), assembled with 0° Hopkins Rod, was introduced through the larynx to evaluate the anatomic condition of the airway, observing the whole respiratory way.

Table 2: Cardiovascular congenital pathology associated with TEF

The aim of the endoscopic evaluation was to assess the presence and define the position of TEF and its cannulation, to verify the patency, before or out of surgery.

Our protocol foresees the use of the endoscopic approach in neonatal age as a preliminary examination before surgical repair of AE in order to classify the type of EA and to verify the presence of unrecognised second upper TEF. The endoscopic approach of suspected recurrent TEF is considered, in infancy, a conservative procedure for the closure of TEF with fibrin glue.

4. Results

In all cases observed, rigid and ventilating airway endoscopy was performed. The tracheobronchoscopic airway evaluation was able to identify the presence, the level and number of TEF in all patients, in order to classify the cases and plan the therapeutic strategy. In three patients (2 EA type II, 1 EA type IV) a second endoscopy was required to locate a fistula of small size in the upper part of the trachea and allow the correct diagnosis of AE. None of the patients submitted to airway endoscopy had any complications related to the procedure.
The endoscopic evaluation showed the fovea of TEF in different positions, in the upper, medium and lower part of the trachea, or in some cases did not detect the presence of fistula (Table 3).

<table>
<thead>
<tr>
<th>Position of TEF</th>
<th>Type of EA</th>
<th>Number</th>
<th>%</th>
</tr>
</thead>
<tbody>
<tr>
<td>Upper TEF</td>
<td>EA II</td>
<td>5</td>
<td>3.5</td>
</tr>
<tr>
<td>Lower/medium TEF</td>
<td>EA III</td>
<td>109 (*)</td>
<td>77.4</td>
</tr>
<tr>
<td>Upper + lower TEF</td>
<td>EA IV</td>
<td>3 (**)</td>
<td>2.2</td>
</tr>
<tr>
<td>Isolated TEF</td>
<td>EA V</td>
<td>5</td>
<td>3.5</td>
</tr>
<tr>
<td>No TEF</td>
<td>EA I</td>
<td>20</td>
<td>13.4</td>
</tr>
<tr>
<td>Total patients</td>
<td></td>
<td>142</td>
<td>100</td>
</tr>
</tbody>
</table>

Table 3: TEF position and classification on the basis of airway endoscopy.

* Recurrent fistula submitted to the closure with thoracic surgery (two cases)
** Recurrent fistula submitted to the closure with thoracic surgery (one case)
≠ Recurrent fistula submitted to the closure with endoscopic conservative therapy (one case)

Figure 1. Tracheoscopy in upper TEF: air bubbles from posterolateral membranous trachea
The fovea located in the upper part of the trachea was always of small size, and difficult to diagnose, communicating with the superior esophageal pouch: in 5 cases an EA of type II was detected, always associated with a flat abdomen and radiologically gasless (Figure 1).

Figure 2. Tracheoscopy in lower TEF: large fovea of membranous trachea
The fovea located in the lower or medium part of the trachea was always of large size, and simple to identify, being located in the back of the trachea at the level of pars membranacea: in our series this is the most common type, communicating with the inferior esophageal pouch and, if single, can be classified in the EA of type III (Figure 2).
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5. Discussion

In our experience the tracheobronchoscopic airway evaluation is a fundamental procedure in the presence of symptoms such as choking, coughing, cianosis at feeding and for the diagnostic and therapeutic management of AE. Endoscopy is in fact the only procedure for the recognition of tracheoesophageal fistula (TEF) and allow a reliable classification.

The literature contains various descriptions of airway endoscopy used for the recognition of TEF, performed both with flexible or rigid instruments (5, 6, 7). The use of the rigid endoscopy...
is reported in numerous studies (2, 8, 9) and has always been our technique of choice. It allows us to assure an open airway and assists operative management: in the presence of TEF the airway endoscopic procedure was in fact diagnostic, and operative at surgery.

From a diagnostic standpoint, the endoscopy allows the identification of the EA type, the characteristics and size of the fistula, as well as the anatomic relationship with the esophageal pouches. From an operative point of view the endoscopy allows the fistula themselves to be incannulated, which, in the case of distal TEF, gives access to the stomach, which can then be drained, reducing the risks of aspirative pneumonia and improving the mechanical ventilation. The identification of the precise anatomic position of the TEF allowed the tip of the endotracheal tube to be adjusted in order to achieve the optimal ventilation. Large or carinal fistula may bring about potential complications during anesthesia if the endotracheal tube happens to slip into them. The presence of the catheter or set of balloon catheters through the fistula prevents gastric insufflation, reducing the risk of pulmonar aspiration, and guides the surgical dissection during minimally invasive or open surgery (10, 11, 12, 13).

In our experience the recognition of an upper fistula is particularly difficult because the fovea is smaller, or not present, compared to lower TEF. This anatomical condition explains the need for a second endoscopy to better identify the presence of the upper fistula and plan correctly the therapeutic strategy: the presence of upper fistula led us to revise the diagnosis of two EA at first considered of type III, to AE of type IV, and one EA of type I without fistula to EA of type II.

In the absence of esophageal atresia, the classic symptoms indicate the rigid endoscopy as the gold standard to exclude TEF; in persisting symptoms after the correction of esophageal atresia, a new endoscopic procedure is indicated for a differential diagnosis of recurrent TEF or to exclude the presence of a second upper fistula.

6. Conclusions

In the presence of evident esophageal atresia, but also when choking, coughing, cianosis at feeding are present, the tracheal endoscopic approach in expert hands is an appropriate diagnostic procedure to apply in neonates and in infancy.

In our experience the rigid ventilating tracheobroncoscopy improves perioperative management of newborns affected by EA and appears a safe and effective procedure.

7. References


