# Unusual presentation of three tracheoesophageal fistulas without esophageal atresia.

Luis Falcón [1, 2]

**Abstract**

Tracheoesophageal fistula without esophageal atresia is a very rare variant, and, much more rarely, it is the presence of three fistulas. Below it is presented the case of a newborn with swallowing problems, who is made a quick and accurate diagnosis of such malformation.

**Key words:** Congenital defect. E-Type fistula. H-type fistula. VACTERL

1. Servicio de Neumología Pediátrica. Hospital de Emergencias Pediátricas. Lima, Perú.
2. Servicio de Neumología. Clínica Internacional. Lima, Perú.

**How to cite the article:** Falcon L. Unusual presentation of three tracheoesophageal fistulas without esophageal atresia, a case report. Interciencia RCCI. 2018; 8(2) 69-72

## Introduction

Tracheoesophageal fistula (TEF) is a congenital malformation with an incidence varying between 1:2500 and 1:3500, and which is usually associated with the presence of esophageal atresia. There are five subtypes, depending on how the esophagus or the trachea is affected, and each subtype has a frequency of presentation[1]. TEF without esophageal atresia (type E) is one of the most infrequent subtypes (1 in 87,000 births[2]).

The etiology is still unknown, but it is known to be based on embryological development. Some mutations have been identified in N-myc, Sox2 and CHD7 which are related to their origin, and TEF are frequently associated with other malformations, such as vertebral, anal, tracheal, esophageal, renal and limb disorders (VACTERL)[3].

This type of fistula is often unperceived, even into adulthood. Bronchoscopy and endoscopy are the most helpful methods for diagnosing TEF[4], and its treatment is mostly successful, with low percentages of complications[5].

## Case

A 31-day-old male patient, from Cuzco, was hospitalized (in that city) from his birth due to swallowing disorder. He had no relevant family or prenatal history. Since he was born, he was given breast milk by an orogastric tube. A radiographic study of swallowing was performed with a contrasted substance and it was noticed the passage of contrast substance from the digestive system to the airway (see Figure 1). He was referred at 6 days old to a hospital in the city of Lima for diagnosis and treatment.

When he was hospitalized in Lima he was awake, reactive, thin, and pale, the rest of the physical examination was normal. In the auxiliary examinations, mild anemia and plateletopenia were found, as well as uncompensated respiratory alkalosis. On the second day of hospitalization, a flexible bronchofibroscopy

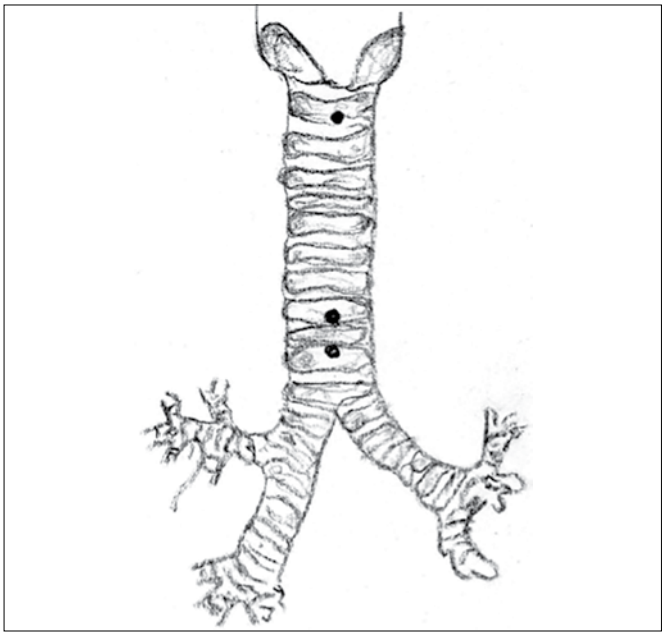

**Figure 1.** Outline of the trachea and location of the fistulas.

was performed and three tracheoesophageal fistulas (TEF) were found: two distal fistulas and one small proximal fistula (see Figure 2). In Multislice CT (day 2 of hospitalization), it was found: i. A TEF of 6 mm in diameter; ii. brachiocephalic trunk in bovine arch (anatomical variable); and, iii. aberrant right subclavian artery. The echocardiogram (day 10 of hospitalization) showed: levocardia, situs solitus, and permeable foramen ovale.

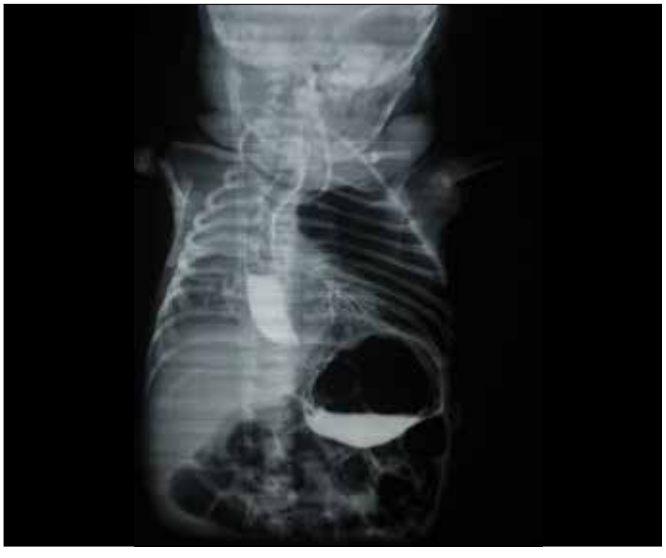

**Figure 2.** Passage of contrast substance from the esophagus to the airway.

The fistulas were treated in two stages. The first intervention was by video-assisted thoracoscopy surgery (14th day of hospitalization), in which the two distal fistulas were closed, separated and transfixing stitches were placed covering both fistulas (see Figure 3). In the second intervention (24th day of hospitalization), a surgery was performed to close the proximal fistula. It was accessed through the anterior triangle of the neck; it was closed with two transfixing stitches (see Figure 4).

During hospitalization, the patient had aspiration pneumonia before the definitive closure of the TEFs. The values of PO2, before the closure of the fistulas, fluctuated between 40 and 50 mmHg and, after closing, reached 97 mmHg.

**Figure 3.** Distal fistulas. Moment of closing both of them.

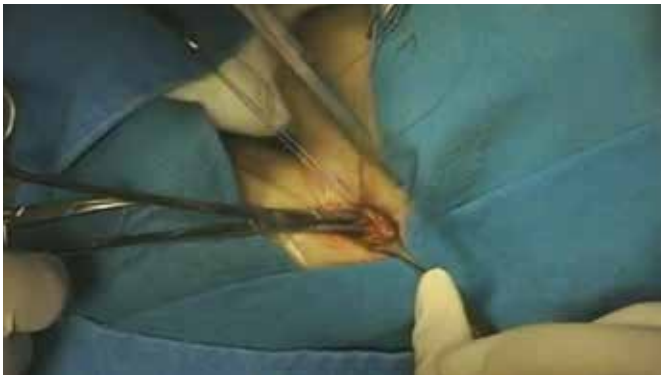

**Figura 4.** Closure of the proximal fistula. Note the access is through the neck.

## Discussion

The presence of TEF without esophageal atresia is rarely frequent, and the presence of three fistulas is even less frequent. There is only one similar report around the world[6].

It was also found an association with other cardiovascular malformations, unlike another series of cases in which association with malformations of the distal digestive system[7] was reported.

This study shows the rapid and accurate diagnosis made by flexible bronchoscopy, followed by surgical repair for the management of an extremely rare case of three tracheoesophageal fistulas, without the presence of esophageal atresia.

The diagnostic evaluation of the TEF should preferably be made through flexible bronchofibroscopy[8]. In this case, the evaluation was almost immediate and superior to the thorax Multislice spiral CT that only reported one fistula. An alternative with very good results, for similar cases, is magnetic resonance[9].

Considering the suspicion of this malformation is essential to carry out a detailed evaluation to have an accurate diagnosis and, thus, to offer an optimal treatment. The management of choice is surgical.

**Help or sources of finance**
None.

**Conflict of interest**
The authors report no conflicts of interest regarding this manuscript.

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**Correspondence:**
Luis Falcón
Unidad de Investigación y Docencia
Clínica Internacional
Av. Garcilaso de la Vega 1420, Cercado de Lima, Perú.
Teléfono: +51 997974262
**E-mail:** lfalconcrest@gmail.com