LUPINE PUBLISHERS

Progressing Aspects in Pediatrics and Neonatology

DOI: 10.32474/PAPN.2023.04.000187

ISSN: 2637-4722

Case Report

A Rare Case of a Variant of Type-C Esophageal Atresia with Double Distal Fistula (IIIb12). Is There a Need of a New Classification?

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Received: May 03, 2023 **Published:** May 12, 2023

Abstract

Esophageal atresia is the most common congenital anomaly of the esophagus. A wide variety of subtypes have been defined in the literature. Kluth's classification is the most complete and descriptive one. To achieve an effective repair, it is vital to perform a tracheobronchoscopy before the operation, to study the anatomy of the respiratory tree and detect tracheoesophageal fistulas. We present the case of a preterm neonate in whom a pre- surgical bronchoscopy showed two distal tracheoesophageal fistulas, making the diagnosis of a type IIIb12 esophageal atresia (Kluth's classification). An end-to-end anastomosis was performed, and we placed a dermal regeneration sheet to help prevent leakages. The surgery was well tolerated, and no major complications were observed. We would like to highlight the importance of performing a tracheobronchoscopy before the surgery. In our case, it allowed us to diagnose a rare case of esophageal atresia, and the complete analysis of the defect's structure led us to a successful correction.

Keywords: Esophageal atresia; tracheobronchoscopy; tracheoesophageal fistula; Kluth's classification; dermal regeneration sheet

Introduction

Esophageal atresia (EA) represents the most common congenital anomaly of the esophagus with an incidence of one in 2,500-4,500 live births [1]. The most widely used classification worldwide is the one described by Gross in 1953, where type C esophageal atresia represents the most common subtype in 80% of cases [2,3]. However, the best description was made by Kluth in 1976, who categorized it into 10 types (Table 1), each having 20 subtypes [4]. It is very important the understanding of the anatomy of the malformation to accomplish a successful repair and for this a pre-surgical bronchoscopy is an essential preoperative exploration. Benjamin [5] first highlighted the importance of preliminary tracheobronchoscopy (TBS) in the management of newborns affected by EA. It is useful for defining the anatomy of the respiratory tree, confirming the presence of tracheoesophageal fistula (TEF), the location and the exact site of entry [6]. We describe a patient with a type C esophageal atresia (IIIb12 according to Kluth's classification) in whom the TBS allowed us to detect the unusual variant of EA such as double distal TEF.

Table 1: Classifications of EA [2-4].

Vogt	Gross	Kluth	Description of Anomaly
1		I	Absent distal esophagus
2	A	II	Pure esophageal atresia
3a	B	IIIa	Esophageal atresia with proximal tracheoesophageal fistula
3b	C	IIIb	Esophageal atresia with distal tracheoesophageal fistula (20 subtypes)
3c	D	IIIc	Esophageal atresia with proximal and distal tracheoesophageal fistula
	G	IV	Membranous atresia
		V	Esophageal atresia with esophageal duplication
		VI	Esophagobronchial communications
4	E	VII	H fistulas
	F	VIII	Esophageal stenosis
		IX	Tracheal atresia or agenesis with tracheoesophageal fistula
		X	Tracheoesophageal fissures or clefts

Case report

We present the case of a 31 weeks' gestation male neonate born to a 32-year-old mother weighing 1594 g. Forty years ago, his father underwent surgery, because of the repair of a type III EA. The parents were not consanguineous. It was a trichorial-triamniotic pregnancy and prenatal ultrasound on the third trimester did not report polyhydramnios. Our patient was born at 31+3 weeks via c-section and was admitted to the Neonatal Intensive Care Unit. He required non- invasive mechanical ventilation during the first hours of life due to mild respiratory distress. The initial physical exam was normal. The diagnosis of EA was made when having difficulties passing an orogastric tube to manage secretions. An X-ray with intra-tracheal contrast was performed (Figure 1), which revealed a proximal dilated esophageal pouch and the presence of air in the stomach, the reason why a TEF was suspected. An 8 Replogle catheter was placed in the upper esophageal pouch to suction secretions. After ruling out major heart malformations, surgical correction was made on the second day of life.

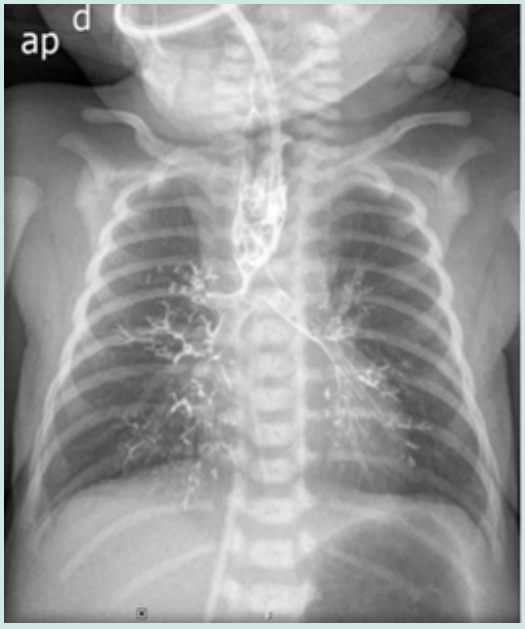

Figure 1: Antero-posterior x-ray and esophagogram with water-soluble agent demonstrating EA. Dilated proximal esophageal pouch. A distal fistula is suspected due to the presence of gas in the stomach.

A pre-surgical bronchoscopy was carried out revealing a double distal fistula, with no signs of tracheomalacia. Both fistulas were located at the level of the trachea and opened 0.5-1 cm above the carina (Figure 2). A right thoracotomy was performed through the 4th intercostal space. We confirmed the presence of a double distal fistula and between them an esophageal segment reduced in its

diameter. We carried out the cutting and closure of the fistulas and an end- to-end esophageal anastomosis with tension after resecting a minimal part of the proximal esophagus and the distal esophageal segment between fistulas. A dermal regeneration sheet (Integra®) was placed between the esophagus and closed TEFs. A transthoracic catheter was left for 24 hours. Trans anastomotic nasogastric feed commenced on the fifth postoperative day. An esophagogram was carried out (Figure 3), on the 12th post-surgical day which showed integrity of the anastomosis without leaks. After almost 3 months our patient remains asymptomatic.

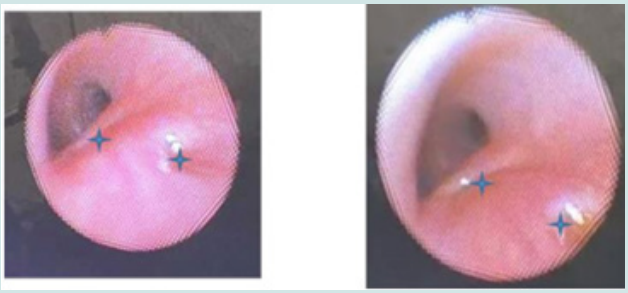

Figure 2: Intraoperative bronchoscopy displaying two distal TEFs.

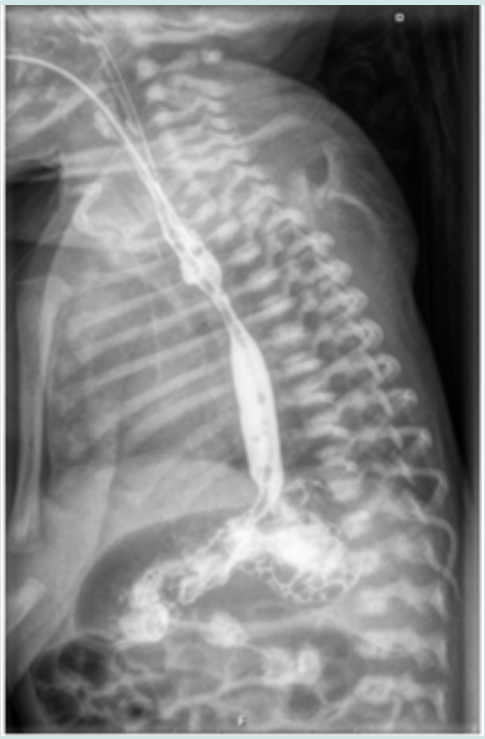

Figure 3: Esophagogram performed on the 12th post-surgical day. No fistulas are observed, with satisfactory passage of contrast through the esophagus to the stomach.

Discussion

Although esophageal atresia is a surgical condition with a high overall survival rate (90%), the prognosis and the success of the intervention depend on the presence of possible associated malformations [7,8]. It is very important to study the anatomy of the esophageal atresia before surgical intervention to achieve a complete and successful repair. Preoperative TBS is the most useful exam in the diagnostic and therapeutic assessment of neonates affected by EA. It provides a more accurate anatomical definition of the anomaly than other diagnostic tools [9-11]. In our case, flexible bronchoscopy allowed the characterization of the two distal fistulas, which might otherwise have gone unnoticed. No absolute contraindications to TBS are reported in neonates. Relative contraindications include pulmonary hypertension and uncorrected bleeding diathesis. Flexible bronchoscopy provides the best assessment for epiglottic collapse, laryngomalacia, tracheomalacia and vocal cord paralysis. Our type of EA is an extremely rare variety and hardly reported in the literature in recent years. Moreover, the use of a biomaterial as an esophagotracheal coverage (Integra®) may represent a good alternative when tissues do not look so healthy. With that, our aim was to prevent leaks, and

we achieved it successfully. Another of the main rarities in our case is the pattern of inheritance, since his father had presented type IIIb EA. Even though certain studies such as the one by Choinitzki et al. suggest that there is no increased risk of AE development in offspring, in recent years it has been revealed that some alteration in the FOX or Shh gene could be associated with the appearance of hereditary AE [12].

Funding

No funding or grant support.

Conflict of Interest

We declare that there is no conflict of interests.

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DOI: 10.32474/PAPN.2023.04.000187