

## A Preoperative Diagnostic Challenge of a Long Overlapping Upper Pouch with Distal Tracheoesophageal Fistula

Takafumi Kondo<sup>1</sup>, Shohei Honda<sup>1</sup>, Masashi Minato<sup>1</sup>, Sorahiko Fujisawa<sup>1</sup>, Hisayuki Miyagi<sup>1</sup>, Kazutoshi Cho<sup>2</sup>, Hisanori Minakami<sup>2</sup> and Akinobu Taketomi<sup>1</sup>

<sup>1</sup>Department of Gastroenterological Surgery I, Hokkaido University Graduate School of Medicine, Sapporo, Japan

<sup>2</sup>Maternity and Perinatal Care Center, Hokkaido University Hospital, Sapporo, Japan

\*Corresponding author: Kazutoshi Cho, MD; Maternity and Perinatal Care Center, Hokkaido University Hospital, Kita-14, Nishi-5, Kitaku, Sapporo, Japan, Tel: +81-11-706-5927; Fax +81-11-717-7515; E-mail: [chotarou@med.hokudai.ac.jp](mailto:chotarou@med.hokudai.ac.jp)

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### Abstract

A wide spectrum of variations can occur in esophageal atresia because of the complexity of tracheoesophageal development. We herein described a 0-day-old boy with type IIIb6 esophageal atresia. Esophagography revealed a long overlapping upper pouch that was similar to esophageal stenosis. Gastrostomy is useful for the therapeutic diagnosis of such a case.

**Keywords:** Esophageal atresia; Type IIIb6; Long overlapping; Esophageal stenosis

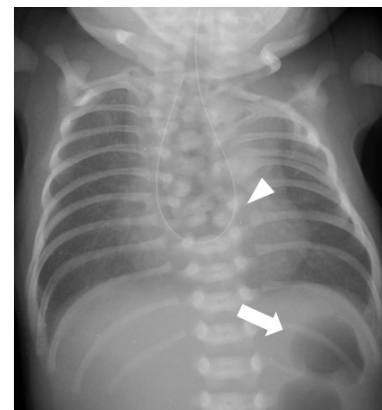
### Introduction

A wide spectrum of variations can occur in esophageal atresia (EA) because of the complexity of tracheoesophageal development. One of the most common classifications of EA is Gross classification, including types A to E. On the other hand, Kluth et al. [1] reported ten large classifications and ninety-six small classifications of EA based on the presence or location of tracheoesophageal fistula (TEF), the gap distance, and the shape of the upper pouch. Kluth type IIIb6 has been defined as esophageal atresia with TEF of the distal segment only and a long proximal dead-end esophagus overlapping with the TEF and distal esophagus. We herein presented a 0-day-old boy with type IIIb6 EA with a long overlapping upper pouch that reached close to the diaphragm. Esophagography revealed that the long overlapping upper pouch was similar to esophageal stenosis, which made it difficult to diagnose preoperatively.

### Case Report

This case involved a 37-week-gestation boy born by an uneventful vaginal delivery with a birth weight of 2,927 g. Polyhydramnios and an absent gastric bubble were detected by fetal ultrasound during gestation. He had combined anomalies such as vertebral body and rib anomalies, and a double outlet right ventricle. A chest radiograph after the insertion of a firm 10 Fr. catheter through the mouth showed a dilated esophageal pouch, the coil-up catheter, and a gastric bubble (Figure 1). We performed bronchoscopy to diagnose EA, but could not identify the orifice of TEF. Esophagography showed that the upper esophageal pouch was dilated close to the diaphragm. The flow of contrast media into the stomach was not detected (Figure 2), and we could not insert a catheter into the stomach. It was difficult to distinguish EA from congenital esophageal stenosis at the lower esophagus from these radiographic findings. Therefore, we decided to conduct gastrostomy under general anesthesia for a therapeutic diagnosis. In retrograde esophagography from gastrostomy, the trachea was visualized with contrast media; therefore, the presence of TEF was

confirmed, resulting in a definitive diagnosis of EA with Gross type C. Primary repair by end-to-end anastomosis of the esophagus was subsequently performed. Intraoperative findings revealed that the blind end of the upper esophageal pouch close to the diaphragm, and usual tracheal bifurcation gave an origin to TEF. The postoperative course was uneventful, the patient could feed independently, and his weight had increased well by the 26th postoperative day.

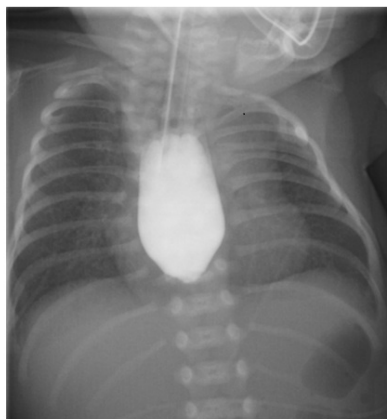


**Figure 1:** Chest radiography showed a dilated esophagus, the coil-up catheter (white arrow head), and a gastric bubble (white arrow).

### Discussion

EA can generally be diagnosed by preoperative imaging, such as chest radiography, the failure to pass an infant-feeding tube into the stomach, or esophagography. On the other hand, Kluth type IIIb6 is an extremely rare variant of EA that is difficult to diagnose preoperatively because the finding of a long overlapping upper pouch is similar to that of esophageal stenosis at the lower esophagus due to the blind end of the upper pouch growing long enough to reach the level of the diaphragm.

Rathod et al. showed that the traditional infant-feeding tube test was unreliable in such a case [2]. Rathod et al. also reported that the presence of a long overlapping pouch may be the reason for a late diagnosis and delayed referral [3].



**Figure 2:** Esophagography showed a dilated esophagus close to the diaphragm.

In present case, we confirmed the diagnosis using retrograde esophagography from gastrostomy during surgery. We failed to detect the orifice of TEF with a bronchoscopy, which gave the false impression of esophageal stenosis. Mahalik et al. and Garge et al. both reported that the use of pre-operative CT scans could not be generalized and protocolized as a standard of care in the management of TEF because the safety of this technique was questionable due to limited facilities and associated radiation hazards [4,5].

Nevertheless, a preoperative CT scan for a rare variant of EA, such as Kluth type IIIb6 in our case, may provide good anatomical delineation and help in surgical decision making.

Gastrostomy is typically performed in cases in which two-stage surgery is required. However, we herein demonstrated that gastrostomy was useful for a therapeutic diagnosis in such a rare variant of EA as the present case, even though one-stage surgery can also be performed.

In conclusion, it is important for pediatric surgeons to be aware that there are some rare variants of EA that are difficult to diagnose preoperatively, and an appropriate assessment of type of EA is crucial for careful and meticulous surgery.

## References

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