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Case Report

Esophageal Atresia of Kluth Type-13: Management of a Rare Variant

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Keywords
Congenital malformation
Esophageal atresia
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Abbreviations
EA - Esophageal atresia
POD - post-operative day
TEF - *Tracheo-esophageal fistula*

Abstract

A full-term low birth-weight female newborn presented with clinical features of esophageal atresia. Investigations revealed Kluth type-13, an extremely rare varient of esophageal atresia without fistula, in which the upper pouch is long and ends blindly just above the diaphragm near the gastro-esophageal junction. She was managed with esophagostomy and feeding gastrostomy in the neonatal period, followed by gastric tube esophagoplasty at 2 years of age. At 18 months of follow-up she is thriving well and asymptomatic.

INTRODUCTION

Esophageal atresia (EA) is a congenital anomaly in which the continuity of the esophagus is disrupted with or without persistent luminal communication with the trachea. Its incidence is approximately 1 in 3500 live births.[1] The most common type is an esophageal atresia (EA) with a distal tracheoesophageal fistula (TEF) (84%), while pure EA is much less common (6%).[1] Pure EA is commonly found in the upper one-third of the esophagus and is characterized by a wide gap between the two atretic ends.

Kluth classified EA into 10 different types with 96 sub-types based on: (i) the presence, location and number of fistulae, (ii) the gap between the atretic ends, (iii) the shape of the upper pouch, (iv) the presence of cyst, stenosis, strands, membranes and duplications, (v) associated abnormalities of the trachea, (vi) broncho-esophageal communication and (vii) esophago-laryngo-tracheal fissure. Kluth type-13 is a rare EA with a long upper esophageal blind pouch and agenesis of the distal esophagus.[2] We report a rare variant of the Kluth type-13 in which the distal esophageal segment was atretic rather than being totally absent.

CASE REPORT

A 4-day-old female neonate, born normally at full-term with a birth-weight of 1.45 kg, presented with drooling of saliva, regurgitation of feeds and non-passage of meconium since birth. She had had intra-uterine growth restriction. On examination, the abdomen was soft with no distention. Infant feeding tube could not be insertion and it coiled in the lower esophagus. Radiographs showed gasless abdomen and blind ending esophagus at the level of the diaphragm. (Fig. 1) Sonography of the spine and urinary system, and a 2D-echocardiography were normal.

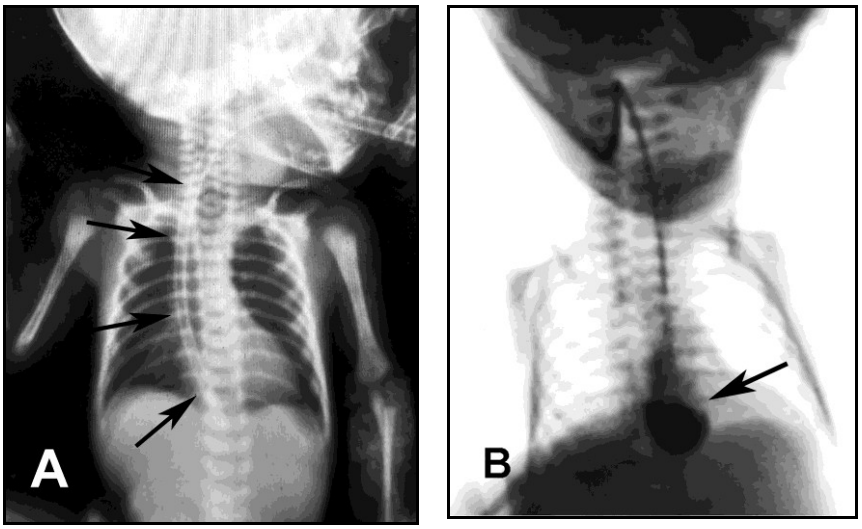

Fig 1. (A) *Plain radiograph showing an esophageal a red-rubber tube in situ (arrows) and a gasless abdomen; (B) Contrast esophagogram showing a long upper esophageal blind pouch that reaches upto the diaphragm (arrow). Gasless abdomen suggests atretic esophagus without fistula.*

She was diagnosed with EA without tracheo-esophageal fistula. After brief resuscitation, a laparotomy was done by a midline incision. The stomach was found to be small. The esophagus was atretic at gastro-esophageal junction with a 0.7-1 cm gap between the blind ends. Stamm's feeding gastrostomy along with a cervical loop esophagostomy were done. An infant feeding tube could not be passed retrogradely into the distal esophagus from the gastric lumen.

Persistent apneic spells during the post-operative period necessitated mechanical ventilatory assistance for 3 days. Feeding through gastrostomy tube was started on the postoperative day-10. After ensuring adequate weight gain, she was discharged on the 16th postoperative day (POD).

She was followed-up regularly as outpatient and a definitive operation was done at 2 years of age. The gastrostomy was reversed by laparotomy. The distal esophageal segment attached to the gastric fundus was atretic and measuring 4 cm in length. (Fig. 2) Gastric tube of 5 cm length was created from the greater curvature of the stomach. (Fig. 2) Tension free esophago-gastric tube anastomosis was done over a trans-anastomotic 12Fr Freka's tube. Oral feeds were started on the POD-5 and the Freka's tube removed on POD-10. She recovered uneventfully and was discharged on the 12th POD. At 18 months of follow-up, she is asymptomatic and thriving well.

DISCUSSION

In 1697, Thomas Gibson reported the first case of EA confirmed at post-mortem examination.[3] The diagnosis of EA is suspected by the inability to pass an infant feeding tube into the stomach due to bind ending of the upper esophagus. The resistance is generally felt at about 10 cm from the lower incisors.[4] If the infant feeding tube passes beyond this distance and then meets resistance, the possibilities of perforation of the upper pouch, a distal web or a stenosis should be suspected.[1] A gasless abdomen on X-ray in case of EA suggests the absence of a distal TEF.[4] Differentials include pure EA or EA with a proximal TEF. Our case was distinct as the patient had atresia of the distal esophagus without fistula.

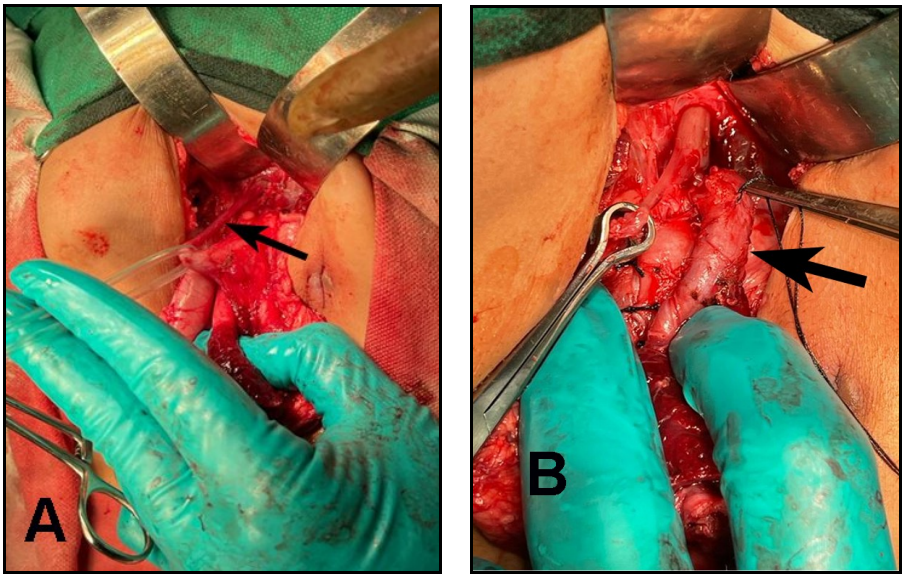

Fig 2. *Intraoperative photographs showing (A) atretic lower segment of the esophagus (arrow) and (B) the reconstructed gastric tube (arrow).*

Various theories have been put to explain EA with or without TEF. One of them holds that the trachea initially grows as a part of the undivided foregut and then becomes an independent structure due to the separation process that starts at the level of the lung buds proceeding in a cranial direction.[5] This is associated with a precise temporo-spatial pattern of expression of the key developmental

It is possible to measure the gap between the two ends of the atretic esophagus in terms of vertebral body heights. This requires insertion of a radio-opaque catheter in the proximal esophagus and injecting radio-opaque contrast or passing a bougie via gastrostomy site into the lower esophagus. Where the gap is less than the height of 2 vertebral bodies, immediate primary anastomosis can be done. Delayed primary repair can be done for a gap of 3-6 vertebral length. For a gap of more than 6 vertebra esophagostomy and gastrostomy should be done.[6]

Kluth classified EA into various sub-types depending upon the anatomical features.[2] The type-13, i.e. long upper esophageal blind pouch with total agenesis of the distal esophagus, is a rare variety of EA. Our case can be a further variant of Kluth type-13 as distal esophagus was atretic rather than being totally absent.

Gupta et.al reported 2 cases of Kluth type-13 EA. One of them was a 2-day-old term neonate 2.2 kg who underwent feeding gastrostomy and esophagostomy and the second one succumbed before any surgical intervention.[7] Shawyer and Flageole reported a case of intra-abdominal EA without TEF, wherein a primary esophageal anastomosis was done by an abdominal approach.[8]

Our patient had a birth weight of 1.45 kg and was at high risk for survival. We did cervical esophagostomy and feeding gastrostomy as a primary lifesaving procedure and scheduled the definitive repair at 2 years of age. We emphasize that all pediatric surgeons must be aware of this rare variant of EA for optimal outcome.

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