Kluth type I\textsubscript{3} and intra-abdominal variants of esophageal atresia: A case series

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ABSTRACT
Background: Esophageal atresia (EA) encompasses a group of congenital anomalies (one in 2500 live births) comprising an interruption in the continuity of the esophagus combined with or without a persistent communication with the trachea. It is confirmed by passing no. 10 sterile, blunt-tipped red rubber catheter into the esophagus, which gets failed to pass beyond 10 cm.

Case Series: We describe two male neonates in whom the infant feeding tube could be passed to 18-20 cm in the upper esophageal pouch. A babygram with a blunt-tipped soft red rubber catheter in situ confirmed the esophageal atresia (EA) with the long upper pouch in the first case and EA with obstruction at the gastroesophageal junction in the second one.

Conclusion: Importance of recognizing rare Kluth variants of EA is stressed. A low threshold for performing a red rubber catheter test is stressed.

INTRODUCTION
Esophageal atresia (EA) encompasses a group of congenital anomalies (one in 2500 live births) comprising an interruption in the continuity of the esophagus combined with or without a persistent communication with the trachea. It is confirmed by passing no. 10 sterile, blunt-tipped red rubber catheter into the esophagus. Failure to pass beyond 10 cm (usually) or failure to negotiate into the stomach (occasionally) is considered diagnostic of EA.[1,2]

If it does not pass beyond 10-13 cm, the diagnosis of EA should be considered and confirmed by radiographs with catheter in situ.[1,2] A red rubber catheter test must be performed to rule out EA as there are multiple fallacies of using IFT especially with long upper pouch EA.[1,2] Kluth (1976) described an atlas of 96 variants of EA with variation from the classical presentation.[3]

We describe our experience with rare Kluth variants of EA where red rubber catheter encountered obstruction below the usual level. Management of these rare variants of EA and a review of literature is presented.

CASE SERIES
Case 1: A 2-day-old term male baby, weighing 2200 grams, presented with excessive salivation and regurgitation after attempted feeds. IPT passed approximately 18 cm giving false impression that the tip is in the stomach, while it remained coiled-up in the long, dilated upper esophageal pouch. A radiograph with the red rubber catheter revealed obstruction at T8 vertebral level; the abdomen was gasless (Fig. 1A, B). Abdominal ultrasounds and baseline blood levels including hemogram and cell counts were normal.

A preoperative diagnosis of EA without tracheoesophageal fistula was made. Feeding gastrostomy and esophagostomy was contemplated. The neonate was resuscitated and preoptimized. Intra operatively, the stomach was small in size and non-dilated which was consistent with findings in EA without fistula. No other intra-abdominal malformation was present. Feeding gastrostomy was performed. Neck dissection revealed elongated, dilated, upper pouch (Fig.1C); cervical esophagostomy was performed. The diagrammatic representation of present case is shown in the Figure 2.
Postoperative contrast study confirmed absence of lower esophageal pouch (Fig.2). Gastrostomy feeding was started on 4th postoperative day. Outcome was favorable; the patient is gaining weight on follow-up. We have planned gastric pull up at the age of 1 year in this baby.

**DISCUSSION**

Kluth divided EA into 10 large classifications and ninety-six small types based on the presence or location and number of TEF, the gap distance, and shape of the upper pouch and presence of cyst, stenosis, strands, membranes, duplications, tracheal abnormalities, esophago-bronchial communication and fisture.[3]

EA without fistula and non-demonstrable distal esophagus is Kluth type I; EA with presence of both proximal and distal segments and without fistula is Kluth type II. Kluth type I is further divided into 7 types from I1 to I7 and type II is further divided into 5 types from II1 to II5 on the basis of anatomical characteristics.[3] The overgrown upper esophageal pouch is extremely rare.[2] EA with extremely long upper esophageal blind pouch with agenesis of distal esophagus is Kluth Type I3; it was reported by Durston in 1670.[3] All the features in case 1 were consistent with Kluth Type I3, as evident by gasless abdomen on preoperative radiographs (Fig.1), absence of proximal tracheoesophageal fistula, and absence of lower esophageal pouch as seen on contrast study (Fig.2). Other variants with long upper esophageal pouch are Kluth type IIIb6 (EA with distal TEF with overlapping of the segments) and Kluth type V5 (long overlapping of segments with sharing of common muscular wall); also, Kluth type VIII2 (lower esophageal web) and Kluth type VIII5 (lower esophageal ring) mimic long upper esophageal pouch.[3]

Membranous atresia is Kluth type IV and is further divided into 7 types from IV1 to IV7 on the basis of associated anomalies, with or without fistulae at various sites.[3,4] Congenital esophageal stenosis is Kluth type VIII and is also divided into 7 types from VIII1 to VIII7.[3] In our second case an incomplete
obstruction was present in the abdominal esophagus suggesting either a perforated web or stenosis at GE junction, as there were few gas bubbles in the lower abdomen. There are only a few reports of intra-abdominal EA described in the literature till date.[5-7] In earlier cases of membranous atresia described in the literature, there was membranous obstruction at cardiac end with gasless abdomen suggesting complete obstruction.[5,6] In another case there was intervening non-patent lumen (like a fibrous cord) between the atretic end and the stomach.[7] These types of intra-abdominal EA have not been described in Kluth's Atlas.[3] These features are suggestive of a new variant of EA, which could be categorized as either Kluth type IV8 or Kluth type VIII8 EA.

Initial management (newborn period) of Kluth type I including our first case (Kluth Type I3) is cervical esophagostomy and feeding gastrostomy (owing to the absence of distal esophageal segment). Definitive operation i.e. esophageal replacement surgery is preferably performed at around the age of 1 year (Indian sub-continent).[8]

Usual approach for management of either of either Kluth type IV or Kluth type VIII EA is by thoracotomy and procedure is carried out depending upon the sub-type. In management of latter case (intra-abdominal EA), abdominal approach is used to identify the anomaly (web, etc.) and perform primary anastomosis.

Author recommends that Pediatricians and Paediatric surgeon should be aware and also must diagnose unusual / rare Kluth variants of EA. A low threshold for performing red rubber catheter test is stressed, especially in geographical areas with high incidence of EA.

To conclude, Kluth type I3 and type IV variants of esophageal atresia are very rare. Importance of recognizing rare Kluth variants of EA by performing red rubber catheter test is stressed, especially in geographical areas with high incidence of EA.

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