We, herein, present an extremely rare case of an esophageal atresia (EA) with distal tracheoesophageal fistula (TEF) with additional perforated membrane at the lower one-third of fistula. A neonate presented with difficulty in breathing and excessive frothing from the mouth. Radiograph with red rubber catheter in situ (obstruction at 10 cm from the gum margins) suggested EA with distal TEF. During thoracotomy, after ligation of fistula, a 6 Fr infant feeding tube was introduced into the distal esophagus which revealed obstruction at the lower one-third. An esophagotomy was performed; a membrane with opening at the center was identified. Following its excision, the esophageal end became dusky necessitating esophagostomy and feeding gastrostomy. A high index of suspicion for membranous obstruction at the lower one-third of fistula should be kept in mind while dealing with EA with distal TEF.

**Keywords:** Esophageal atresia, Kluth type IIIb, membranous obstruction, perforated membrane, variant

**INTRODUCTION**

Esophageal obstruction in the newborn is usually related to esophageal atresia (EA).[1] EA with distal tracheoesophageal fistula (TEF) being the most common type is classified as Vogt type IIIb and also Kluth type IIIb.[2] We, herein, present an extremely rare case of an EA with distal TEF with additional perforated membrane at the lower one-third of fistula necessitating esophagostomy and feeding gastrostomy.

**CASE REPORT**

A 4-day-old, low birth weight (2000 g) female neonate presented to us with difficulty in breathing and excessive frothing from the mouth. The patient was premature with 36 weeks’ gestational age and second in birth order; there was no history of congenital malformations in the family. On examination, the general condition of the patient was poor with respiratory rate – 60/min, pulse rate – 120/min, and SpO₂ – 70%. There was mild abdominal distension. After oropharyngeal suctioning, a soft, 10-Fr red rubber catheter was passed in the esophagus but met with an obstruction at 10 cm from the gum margins, suggesting the presence of EA. A radiograph with red rubber catheter in situ suggested EA with distal TEF [Figure 1]. Right posterolateral extrapleural thoracotomy was performed through the fifth intercostal space. External discontinuity of esophagus and anatomy of upper esophageal pouch were demonstrated. The presence of distal TEF was also confirmed; it communicated with trachea 1 cm above its bifurcation. The fistula was transfixxed and ligated. A soft tube (shunt tubing for hydrocephalus surgery) was introduced to confirm the distal patency of esophagus. It revealed obstruction at the lower one-third of the esophagus. It was followed by a smaller caliber 6 Fr infant feeding tube (IFT) which confirmed obstruction. An esophagotomy was performed; a membrane with opening in the center was identified [Figure 2]. Following its excision, the esophageal end became dusky necessitating esophagostomy and feeding gastrostomy. The distal end of the esophagus was ligated before performing feeding gastrostomy. There were no postoperative complications. The patient is on regular follow-up and is gaining weight.

**Address for correspondence:** Dr. Rahul Gupta, Department of Paediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India. E-mail: meetsurgeon007@gmail.com

This is an open access journal, and articles are distributed under the terms of the Creative Commons Attribution-NonCommercial-ShareAlike 4.0 License, which allows others to remix, tweak, and build upon the work non-commercially, as long as appropriate credit is given and the new creations are licensed under the identical terms.

For reprints contact: reprints@medknow.com

**How to cite this article:** Gupta R, Singh A, Gupta AK. Kluth type IIIb, variant of esophageal atresia. J Indian Assoc Pediatr Surg 2020;25:248-50.
Discussion

Kluth in 1976 divided EA into 10 large classifications and 96 small types based on the presence or location and number of TEF, the gap distance, and shape of the upper pouch and the presence of cyst, stenosis, strands, membranes, duplications, tracheal abnormalities, esophago-bronchial communication, and fissure.\[1,2\]

EA with distal TEF is Kluth type IIIb. It is the largest subgroup being further divided into 20 types from IIIb\(1\) to IIIb\(20\) on the basis of anatomical characteristics of upper pouch and fistula and also associated anomalies.\[2\]

We believe that our case is comparable to that described by Tuqan in 1962\[3\] and also Koop and Hamilton in 1968.\[4\] It was classified by Kluth as type IIIb\(9\) in his Atlas,\[2\] but with a small difference that in our case, (a) the perforated membrane was present in the distal end of the esophagus, in contrast to stricture in Kluth type IIIb\(9\), and (b) external wall of distal esophagus in our case was inconspicuous as against obvious narrowing externally in earlier cases.\[2-4\] Thus, our present is a variant of Kluth type IIIb\(9\) EA which is extremely rare.

In case of Tuqan, the presence of distal obstruction was diagnosed intraoperatively, but its confirmation as circumferential stricture was done at autopsy.\[3\] In one recent report, the diagnosis of stricture was made preoperatively and confirmed intraoperatively with the use of IFT.\[5\] In another large study of 225 patients, congenital esophageal stricture was reported in 8% of patients with TEF and 6.67% of cases with Kluth type IIIb EA.\[6\]

Gasless abdomen on plain radiography suggests complete obstruction of the esophagus, while normal gas pattern shows perforated membrane as seen in our case. Preoperative esophago-bronchoscopy helps to identify proximal fistula and rule out web.\[3\] Filston et al. have recommended routine preoperative endoscopy in all neonates with EA.\[7\] Obstruction occurring at the level of the fistula may be treated without its presence being known, since the surgical treatment of fistula may include the resection of the section of esophagus at that level.\[3\] Obstruction distal to fistula may remain unmasked and untreated, thus leading to lethal complications, such as regurgitation and aspiration pneumonia.

Authors recommend that an IFT must be negotiated into the distal esophagus to (a) confirm its distal patency, (b) rule out obstruction at the lower end of esophagus, and (c) decompress the stomach. The level of obstruction indicated by IFT dictates the approach for excision of membrane, either by surgery or endoscopic method or after performing gastrostomy.\[1,8\]

An esophagotomy incision is made after identification of the obstruction by already placed catheter in the esophageal lumen or by noting the change in caliber of the esophagus.\[1\] Excision of the membrane and closure is done transversely (as attempted in our case). In our case esophagostomy and feeding gastrostomy was performed because (a) lower end become dusky intraoperative following excision of membrane, (b) risk associated with double anastomosis, and (c) presence of associated comorbidities.

A high index of suspicion for membranous obstruction at the lower one-third of fistula should be kept in mind while dealing with EA with distal TEF. Authors recommend the use of IFT to rule out obstruction at the lower end of esophagus and to confirm its distal patency.

Acknowledgment

We are sincerely thankful to the Department of Paediatric Surgery, SMS Medical College, Jaipur, Rajasthan, India, for helping in our endeavor.
Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

REFERENCES
1. Gupta R, Sharma P, Shukla AK, Mehra S. Kluth type IV membranous esophageal atresia at middle one-third of esophagus: An extremely rare entity. J Indian Assoc Pediatr Surg 2017;22:254-6.
2. Kluth D. Atlas of esophageal atresia. J Pediatr Surg 1976;11:901-19.
3. Tuqan NA. Annular stricture of the esophagus distal to congenital tracheoesophageal fistula. Surgery 1962;52:394-5.
4. Koop CE, Hamilton JP. Atresia of the esophagus: Factors affecting survival in 249 cases. Z Kinderchir 1968;5:319-29.
5. Sheth NP. Esophageal atresia and tracheoesophageal fistula with distal esophageal stenosis: Preoperative diagnosis. J Pediatr Surg 2008;43:941-2.
6. Newman B, Bender TM. Esophageal atresia/tracheoesophageal fistula and associated congenital esophageal stenosis. Pediatr Radiol 1997;27:530-4.
7. Filston HC, Rankin JS, Grimm JK. Esophageal atresia. Prognostic factors and contribution of preoperative telescopic endoscopy. Ann Surg 1984;199:532-7.
8. Ramesh JC, Ramanujam TM, Jayaram G. Congenital esophageal stenosis: Report of three cases, literature review, and a proposed classification. Pediatr Surg Int 2001;17:188-92.