PST | PEDIATRIC SURGERY IN TROPICS

ESOPHAGEAL ATRESIA, DUODENAL ATRESIA AND ILEAL PERFORATION IN A 1000g PREMATURE NEONATE COMPLICATED BY A RECURRENT TRACHEOESOPHAGEAL FISTULA: A CASE PRESENTATION

Rainier Bence, Mohamed Abdelsalam, Corné de Vos

Division of Pediatric Surgery, Tygerberg Hospital, Stellenbosch University, Cape Town, South Africa

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Keywords	**Abstract**
Esophageal atresia; Duodenal atresia; Tracheoesophageal fistula	**Introduction:** Esophageal atresia (EA) is a common congenital malformation of the upper gastrointestinal tract caused by failure of separation or incomplete foregut development. This rare congenital disease has known associated anomalies ranging from single to multiple congenital anomalies (MCA) including several known chromosomal abnormalities. A combination of duodenal atresia (DA) and EA in infants has been reported in 3 to 6% of EA cases. Added complications of gastric perforations, and even more rare intestinal perforations, make the management of these infants challenging and should be tailored to each patient's clinical status. Despite advances in surgical techniques, complications such as recurrent fistula or stricture formation remain a significant challenge. The rate of fistula recanalization after surgical repair has an estimated incidence of 5%. The known increase in mortality and morbidity of extremely low birth weight (ELBW) infants together with the presence of multiple congenital abnormalities lead to a high complication rate for these complex groups of EA patients. **Case Presentation:** We report a single case of a 1000 g neonate born at

	30 weeks gestation, with a long-gap EA and a distal TEF, a duodenal atresia and cecal perforation that was later complicated by a recurrent TEF. **Conclusion:** This case highlights the importance of early diagnosis, optimizing surgical technique, prompt recognition of postoperative complications and a multidisciplinary approach to managing these complicated congenital conditions.
Abbreviations	EA: Esophageal Atresia MCA: Multiple Congenital Anomalies DA: Duodenal Atresia ELBW: Extremely Low Birth wight TEF: Tracheoesophageal Fistula g: Gram PDA: Patent Ductus Arteriosus ASD: Atrial Septal Defect TPN: Total Parental Nutrition

INTRODUCTION

Esophageal atresia (EA) with a distal tracheoesophageal fistula (TEF) is a common congenital malformation of the upper gastrointestinal tract caused by the failure of separation or incomplete development of the foregut. [(1)] With an estimated worldwide prevalence ranging from 1 in 2,500 to 1 in 4,500 births, this rare congenital disease has known associated anomalies ranging from single to multiple congenital anomalies (MCA) as well as chromosomal abnormalities. [(2)] A combination of duodenal atresia (DA) and EA in infants has been reported in 3 to 6% of EA infants. [(3)] Added complications of gastric perforations, and even more rare intestinal perforations, make the management of these infants challenging and should be tailored to each individual patient's clinical status. [(3,4)]

Most patients born with this complex congenital disease require surgical intervention shortly after birth. [(5)] Despite advances in surgical techniques, complications such as recurrent fistula or stricture formation remain a significant challenge. [(6)] The rate of fistula recanalization after surgical repair has an estimated incidence of 5%, according to large cohort studies. [(7)] The known increase in mortality and morbidity of extremely low birth weight (ELBW) infants together with the presence of multiple congenital abnormalities lead to a high complication rate for this complex group of EA patients. [(3)] Despite advances in risk assessment models, these findings suggest that the Spitz classification retains prognostic significance as a predictor of

mortality and may still inform risk-based patient management strategies in contemporary clinical practice. [8]

We report a single case of an ELBW neonate born with a long-gap EA with a distal TEF, duodenal atresia and cecal perforation, complicated by a recurrent TEF.

CASE REPORT

A 1-day-old 1000-gram (g) baby girl, born prematurely at 30 weeks gestation, presented to our unit with respiratory distress, abdominal distension and frothing from the mouth. Radiographic investigation raised suspicion of DA and EA with a distal TEF. (Fig 1) Clinically the neonate had no dysmorphic features, with a single hemangioma present above her upper lip. An initial echocardiogram revealed that she had a patent ductus arteriosus (PDA) with bidirectional shunting and a large atrial septal defect (ASD) with left to right shunting. Renal ultrasound was normal.

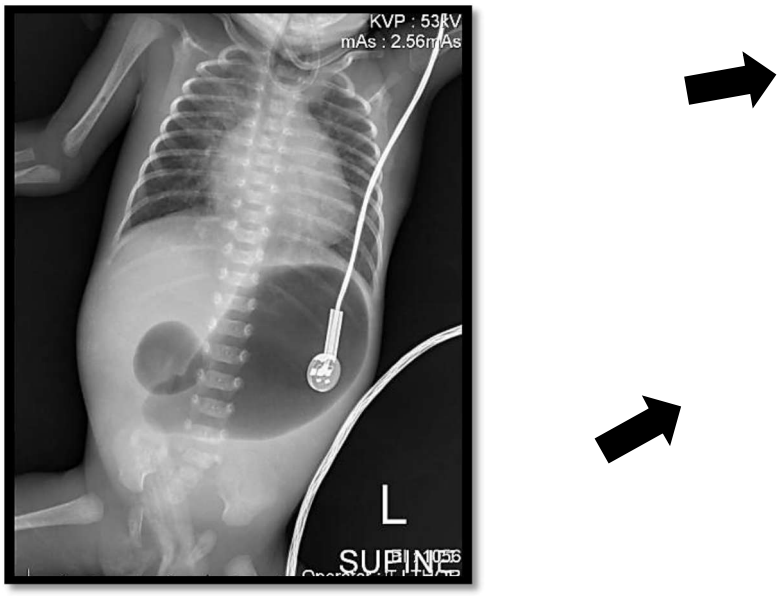

Fig 1. Chest and abdominal radiograph revealing curled NGT and a double bubble sign

The patient was stabilized and taken to theater on day 3 of life. She underwent a right thoracotomy with ligation of the TEF. Due to the instability of the patient, no attempt was made to do a primary esophageal repair at this stage. The TEF was proximal and distally ligated and transected. We proceeded to do an exploratory laparotomy where a Type 3 DA was confirmed and a subsequent cecal perforation found. The cause of this distal perforation was unclear with

no transition zone present but with local contamination. A Duodenoduodenostomy together with resection of the cecal perforation and fashioning of an ileostomy was performed.

Due to the low weight of the patient, the impression of a high proximal esophageal pouch and long gap EA on radiograph together with the presence of MCA, a decision was made to fashion a gastrostomy and left cervical esophagostomy on day 12 of life and not attempt repair of the esophagus. She was intubated and admitted to the neonatal intensive care unit. The postoperative course was initially uneventful, and gastrostomy feeds were slowly being built to full feeds.

At 8 weeks of life the patient suddenly developed respiratory distress with recurrent episodes of coughing, choking, and desaturation. A chest radiograph revealed features suggestive of a recurrent TEF with air visible from the tracheobronchial tree to the stomach. (Fig 2) The patient was stabilized and taken back to theatre for a redo-thoracotomy, redo-fistula ligation and transection of the recurrent TEF.

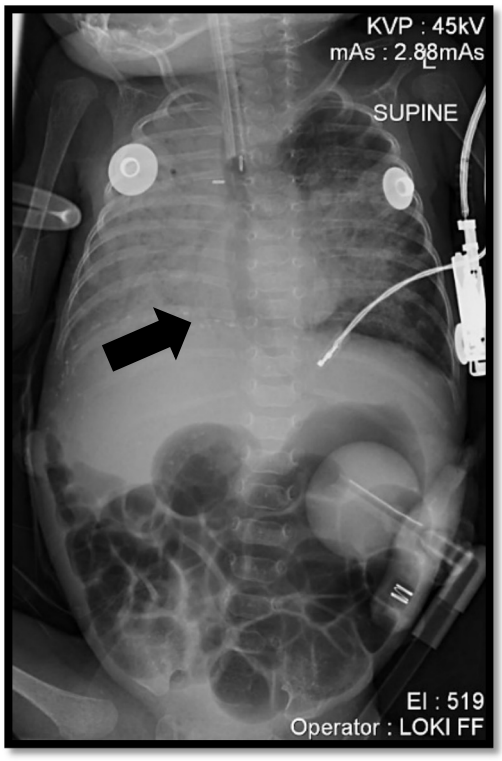

Fig. 2 Chest radiograph demonstrating air in esophagus, suggestive of fistula recurrence

An open right posterolateral trans pleural approach was performed through the previous surgical scar. Adhesiolysis between the esophagus and trachea was released and the fistula was identified. With sharp dissection, using tissue scissors, the fistula was delineated and injury to the surrounding tissue avoided. The fistulous communication was confirmed by the presence of a feeding tube, and the fistula was divided, and both the esophageal and tracheal ends were sutured separately with absorbable sutures. Post-procedure, the patient was initially kept nil per gastrostomy and received continuous total parenteral nutrition (TPN) for the first 14 days. Feeds were slowly re-started via the gastrostomy on day 5 postoperative and increased over the next couple of days after which the TPN were tapered. Repeat echocardiogram three weeks later revealed that both the PDA and ASD had closed.

A subsequent ileostomy closure was performed when she was 6 months old and weighed 1.5 kg. The patient was investigated for growth failure and malabsorption as an underlying etiology. She was referred to genetics for genetic testing and infectious diseases for additional work-up. Gastrostomy feeds were re-started on day 5 post ileostomy closure and full feeds were achieved on day 7.

Although she originally did well post closure, she developed a multidrug resistant *Klebsiella Pneumoniae* bacteremia, had unexplained hypoglycemic attacks secondary to the ongoing sepsis and sadly demised 6 weeks later.

DISCUSSION

EA and DA are rare associated anomalies that does occur in 3 – 6% of these patients. [(7)] The combination of these anomalies makes the management options challenging with a high rate of post-operative complications. We presented a rare case where the patient not only had EA and DA but also ELBW and an unexplained cecal perforation.

The Spitz Classification has been the most widely used prognostic classification for EA patients stating that babies with birth weight of < 1500g have a worse outcome. [(9)] Okamoto et al. revisited this classification in 2009, suggesting that 2000g was an appropriate cut off, much higher than the weight of our patient. [(9)] They suggested that low birth weight with the presence of cardiac anomalies lead to a significant poor prognosis for EA patients.

The most common complication post EA-DA combined repair is gastroesophageal reflux with a significant number of these patients' requiring a fundoplication. [3] A literature review conducted in 2023 included 66 papers and 195 patients born with EA and associated DA. [3] The majority (76.9 %) of patients included in this review presented with an EA and a distal TEF and 61% were premature neonates with an average birth weight of 2160 grams. [3] Our case was consistent with these findings being born premature and diagnosed with an EA with a distal TEF but had a lower birth weight (1000g vs 2160g) compared to their cohort.

Recurrent TEF following primary repair is a well-documented complication of EA surgery. [10] The etiology of recurrent TEF is multifactorial and includes tension on the anastomosis, infection, and ischemia. [6] Redo-surgery for recurrent TEF, while effective, carries significant risks, particularly in neonates with compromised respiratory status. [11] Although open thoracotomy has always been the standard of care, more recently endoscopic management of recurrent TEF has emerged as a less invasive option, demonstrating encouraging results. [7,12-13] To reduce the likelihood of a second recurrence during repair, the following steps should be executed: inserting a catheter through the fistula prior to opening the chest, fully separating the trachea from the esophagus before dividing the fistula, performing rotation-pexy of the esophagus, and placing viable tissue between the suture lines. Our case is consistent with recent studies which suggest that open thoracotomy yields outcomes comparable to endoscopic techniques in the management of recurrent TEF. [13,14]

The successful management of this complication highlights the importance of a multidisciplinary team approach, including pediatric surgeons, radiologists, and anesthesiologists, in achieving optimal outcomes. [6]

CONCLUSION

EA is a rare disease that can be complicated by low birth weight, prematurity and associated with congenital anomalies. Treatment of these patients should be tailored to the individual patient, taking all these factors into account.

Recurrence of tracheoesophageal fistula (TEF) following primary repair remains a challenging complication in pediatric surgery. Awareness of risk factors, such as low birth weight, should

prompt heightened vigilance for potential postoperative complications. Meticulous surgical techniques and individualized postoperative care are essential for achieving favorable outcomes in these cases. This case underscores the importance of early diagnosis, optimization of surgical technique, prompt recognition of postoperative complications, and a multidisciplinary approach in the management of these complex congenital anomalies. a Future directive would be to develop an algorithm to assist in such a rare condition.

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