

REVIEWS

Open Access



Isolated distal esophageal atresia in neonates: a case report and review of the literature

Rasem Keilani¹ , Eman Badran² , Hanan Mansour^{3*} , Rasha Alatout³ and Abdallah Alabadla³

Abstract

Esophageal atresia occurs in 1 out of 2500 to 4500 live births; an isolated variant occurs in 8% of the cases. In this case report and literature review, we present a rare case of isolated distal esophageal atresia happening just 2 cm above the diaphragmatic right leaflet with the connecting band between the two esophageal parts. The location raised the possibility of a direct vascular accident to that segment as a cause of the atresia. Post-operative ambu bagging may facilitate leak at the anastomosis site and should be done carefully, as well as, to introduce contrast with suckling not with jet injection for the same reason. We mentioned that to raise a wariness as this mishap may cause problems.

Keywords: Neonate, Gastrointestinal tract obstruction, Rare isolated esophageal atresia, Distal esophagus

Background

Esophageal atresia (EA) is the most common congenital anomaly of the esophagus [1]. It can be isolated or associated with a proximal or distal tracheoesophageal fistula (TEF). This anomaly occurs in 1 out of 2500 to 4500 live births [2]. Many anatomical variants are described in literature for this anomaly, the isolated EA without TEF (Vogt II, Gross A) accounts for 8% of all cases of EA [2].

Isolated esophageal atresia represents a surgical urgency, that once recognized requires prompt intervention. The diagnosis of EA is suspected antenatally by the presence of polyhydramnios, at birth by excessive salivation and frothy secretions coming from the mouth, and failure to pass a nasogastric tube (NGT) into the stomach. Definitive diagnosis is established by demonstrating coiling of NGT in the chest and absent gastric bubble in a chest X-ray (CXR) [3]. Routine use of esophagogram or pre-op computed tomography (CT) to diagnose EA is not recommended [4].

Primary surgical repair is the preferred treatment modality [5]. A right-sided posterolateral extra-pleural thoracotomy, performed through the fourth intercostal space is the preferred treatment approach. A left-sided thoracotomy is done when a right-sided aortic arch is identified before operation. A thoracoscopic approach can be done based on the surgeon's experience [6].

Our case report presents the birth of a female at our hospital, diagnosed with isolated EA but does not resemble the typical location of the lost segment of esophagus described in literature, but a lost segment in the distal part of the esophagus. We also reported the complications that followed its surgical repair, the recovery of the infant, and a review of related literature. Literature review was done using the keywords: esophageal atresia, distal esophagus, neonatal proximal gastrointestinal obstruction. The case report has been reported in line with the SCARE Criteria [7].

Case presentation

A female who was 38 + 1 weeks gestational age was born at our hospital in November 2021. She was a product of vaginal delivery, with a birth weight of 2200 g, and an APGAR score of 8 and 9 at 1 and 5 min, respectively. The

*Correspondence: hanan.ju@gmail.com

³ Jordan University Hospital, Amman, Jordan

Full list of author information is available at the end of the article

mother was diagnosed with polyhydramnios antenatally, her pregnancy was uneventful, and she is a 27-year-old, healthy lady.

After delivery, the patient was admitted to NICU due to respiratory distress, which improved on continuous positive airway pressure therapy. Later on, examination of the neonate showed stable vital signs, afebrile, drooling frothy oral secretions (Fig. 1), good airway entry bilaterally, abnormal heart sounds, scaphoid abdomen, normal umbilical cord, patent anus, and normal external female genitalia. CXR showed coiled NGT in the chest and absent gastric bubble (Fig. 2).

A diagnosis of isolated esophageal atresia without fistula was suspected. So, the patient underwent screening for VACTER/VACTREL syndrome, which showed a small atrial septal defect, large ventricular septal defect, negative skeletal survey, normal anus, passed meconium in the first day after birth, and normal renal ultrasound. An upper gastrointestinal contrast study was requested (Fig. 3). It showed dilated upper esophagus and hold up of contrast in the chest, suggestive of esophageal atresia.

An abdominopelvic CT scan with intravenous contrast was requested (Fig. 4). It showed markedly dilated cervical and thoracic esophagus down to the level of thoracic vertebra number 9, with no evidence of TEF.

By this we identified a rare type of isolated EA, occurring in the distal part of the esophagus, just above the gastroesophageal junction (GEJ).

The patient underwent repair of EA, through a right posterior-lateral thoracotomy approach; incision was done at the level of the 5th intercostal space due to the distal location of the defect; a gap length of 3 cm long was identified between the two ends of the esophagus; and the proximal end was significantly dilated and long, connected to the collapsed distal end 2 cm long just

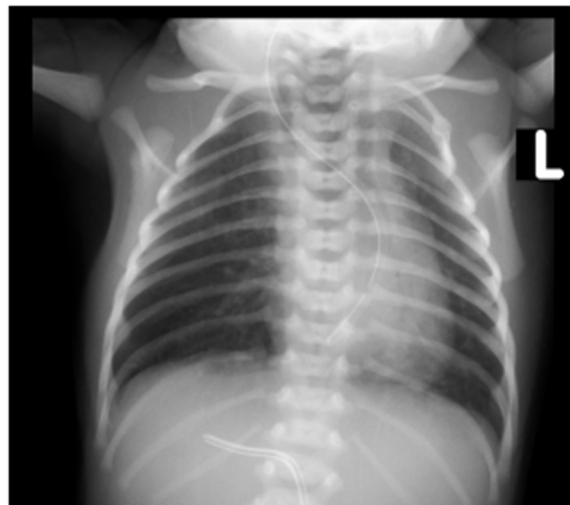


Fig. 2 Coiled NGT in the chest and absent gastric bubble



Fig. 1 Frothy oral secretions and scaphoid abdomen



Fig. 3 Upper GIT contrast study showing the blind-ending esophagus

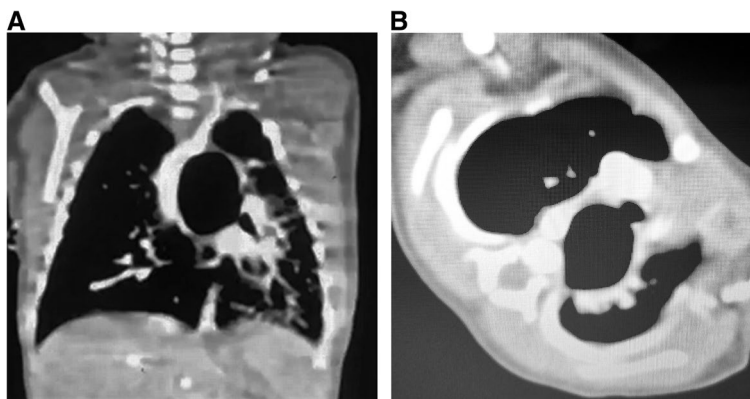


Fig. 4 CT chest with IV contrast; **A** coronal section and **B** horizontal section

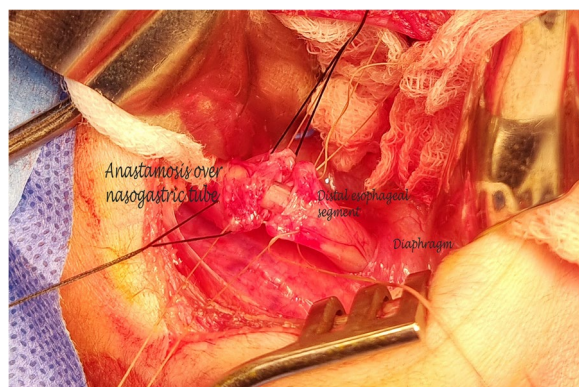


Fig. 5 Intra-op view of cord connection between two ends of the esophagus

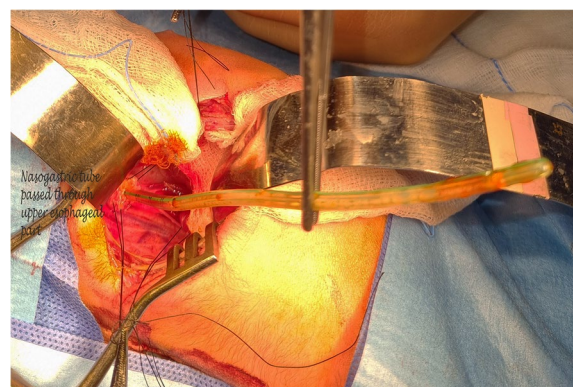


Fig. 6 NGT passing through the proximal esophagus

above the diaphragm by an atretic band. The band was removed; end-to-end primary anastomosis over nasogastric tube using multiple 5/0 Vicryl stitches was done; due to the good length of the upper pouch, no dissection was needed. A right-sided aorta was identified and avoided throughout the procedure as shown in Figs. 5, 6, and 7.

Postoperative days 1 and 2 the patient was doing well, she was maintained on a mechanical ventilator with sedation, serial X-ray post-op showed aeration of the GIT. Thereafter the patient developed multiple episodes of desaturation due to a clogged endotracheal tube (ETT) and frequent ambu bagging. The ETT was changed twice. Follow-up CXR (Fig. 8) showed trapped pneumothorax at the base of the right lung which was managed conservatively.

Upper GI contrast study was done at postop day 8 (Fig. 9), contrast was introduced with pull back technique of the NGT and jet injection of the contrast. It showed a minor leak of contrast from the site of anastomosis. The

patient at the time was still intubated, on a mechanical ventilator, and had stable vital signs. Labs showed no rise of inflammatory markers. She was managed conservatively by keeping her NPO and on total parenteral nutrition (TPN), broad-spectrum antibiotics, and respiratory supportive care.

After 1 week, the upper GI contrast study was repeated, it showed no leak, so the patient was started on feeding orally, she was extubated, and her chest tube was removed.

Before discharge from the NICU, a Gastrografin swallow was done with suckling of contrast material from milk bottle. It showed dilatation of the esophagus with transient hold-up of contrast material at the level of GEJ, and radiological suspicion of stricture at the site of anastomosis (Fig. 10). The stricture was assessed by our pediatric gastroenterologist, who labeled it as not clinically significant. The patient was continued on breast milk feeding orally. She was discharged home at the age of 50 days on Lasix due to her VSD as per cardiology recommendation. She underwent upper endoscopic



Fig. 7 Intra-op image showing primary anastomosis using Vicryl 5/0 sutures

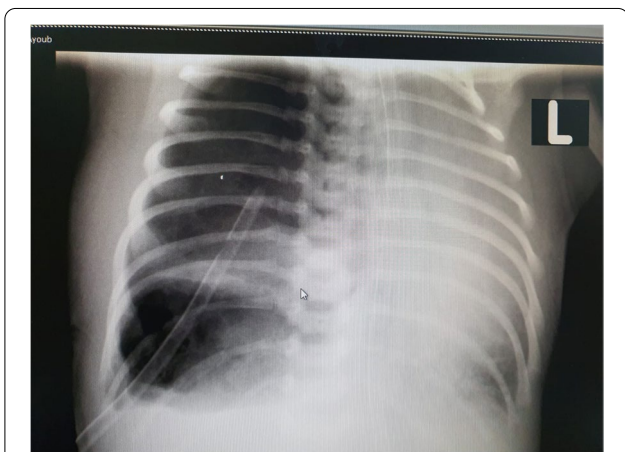


Fig. 8 CXR with trapped pneumothorax and right chest tube in situ

balloon dilatation at the age of 2.5 months and showed good tolerance to feeding after the procedure. The infant is still having failure to thrive type 2, anemia, and vitamin

D deficiency which is followed by her pediatrician in the out-patient clinics.

Discussion and conclusions

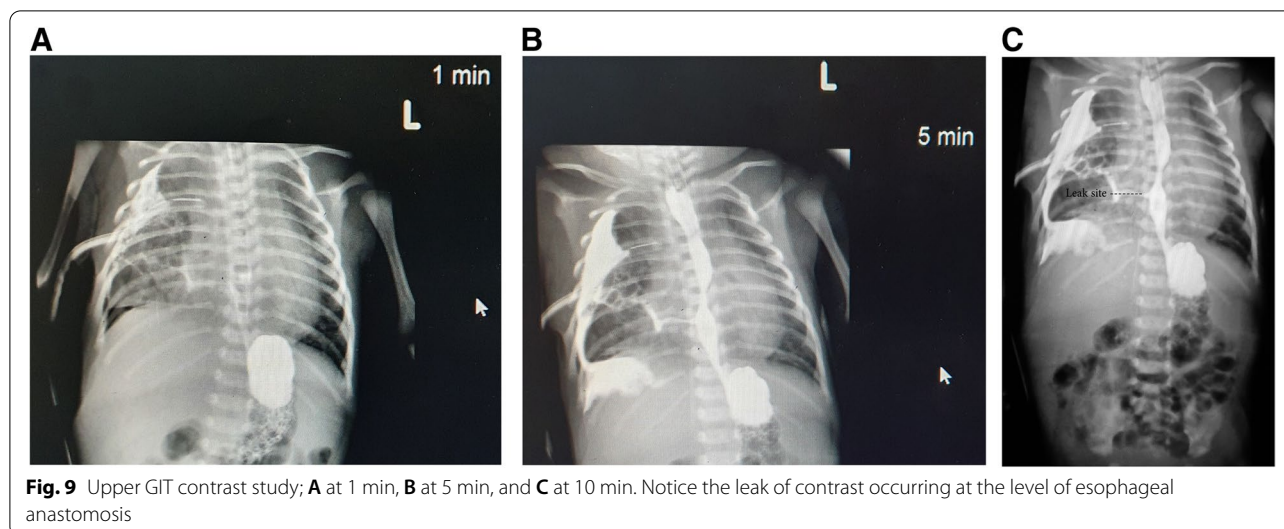
This case demonstrated the occurrence of a special type of isolated EA without TEF, with a peculiar location of the atretic segment occurring just 2 cm above the diaphragm. This has impacted the diagnostic approach and the surgical approach as we needed to enter the thorax a level below the usual level of entry to correct this pathology [6].

The current theories made to explain the occurrence of esophageal atresia, suggesting cellular rearrangement or regression of the primitive digestive tube towards the embryo [2] cannot explain the occurrence of EA in this patient due to the peculiarity of the defect location and the presence of an atretic band. We propose another mechanism related to a vascular accident in-utero that resulted in the loss of a segment of the distal esophagus, like what occurs in type II intestinal atresia [8], we base our theory on the segmental nature of blood supply to the distal esophagus that comes as direct branches from the thoracic aorta [9].

Our patient underwent an upper GIT contrast study which showed a long-dilated esophagus with transient hold-up of contrast just above the diaphragm, then a CT scan to identify esophageal atresia and rule out the presence of a proximal TEF due to the strange location of the atresia, as well as to exclude external pressure on that area by a mass; esophageal duplication or presence of tracheobronchial cartilage remnant in the wall of the esophagus. Recent studies propose the replacement of upper GIT contrast study with saline-aided ultrasonography, aiming to decrease radiation exposure of the infant and its associated risks [10]. The use of pre-op CT scan for surgery planning is controversial and not standardized [4]. The TEF could be missed in CTs in up to 20% of the patients [11].

The complications that followed this surgery are common, occurring in 15–20% of the patients [3]. Leaks are either minor or major, minor leaks are managed conservatively with drainage, prolonged fasting and TPN, and broad-spectrum antibiotics until the leak seals by itself [12, 13]. Some studies compared conservative management with early surgical interventions to close the defect, but no significant difference has been detected [14].

The leak detected in this case could have happened due to frequent ambu bagging in the early post-operative period, which could have weakened the anastomosis site due to the raised intraluminal pressure. This weakness could also explain the occurrence of trapped pneumothorax at the right lower lung zone, which is just opposite to the site of anastomosis.



A retrospective study was done in Beijing Children's Hospital aiming to identify risk factors for pneumothorax after esophageal atresia repair, including 188 patients with Gross type C EA/TEF. They found that postoperative anastomotic leakage and mechanical ventilation were independent risk factors for pneumothorax after EA/TEF repair [15].

Our patient had a minor leak detected on an esophagogram done on postoperative day 8. Despite the leak detected, the patient was afebrile with negative inflammatory markers. Some studies advocate against the routine use of esophagogram postoperative in the absence of predictors of leak [16]. The esophagogram for our patient was done with pullback of NGT that was inserted intraoperatively and jet injection through it with contrast material. We recommend against the use of this technique to do an esophagogram to detect leaks post-op and advocate the administration of contrast by suckling when the infant can swallow saliva.

The presence of stricture post esophageal atresia repair is common, accounting for 45% of the patients [17], and has a specific appearance that can be detected on an esophagogram [18]. Strictures usually respond to a single session of balloon dilatation, and patients show significant clinical improvement in regards of tolerance to feeding and weight gain [17].

In conclusion. This case demonstrated the occurrence of a special type of isolated EA without TEF, with a peculiar location of the atretic segment occurring distally just 2 cm above the diaphragm.

Abbreviations

EA: Esophageal atresia; TEF: Tracheoesophageal fistula; NGT: Nasogastric tube; CT: Computed tomography; NPO: Nil per mouth; GIT: Gastrointestinal tract; CXR: Chest X-ray; ETT: Endotracheal tube; VSD: Ventricular septal defect; NICU: Neonatal intensive care unit; TPN: Total parenteral nutrition; GEJ: Gastroesophageal junction.

Acknowledgements

The authors thank Miss Hadeel Al-Qaisi for editing the English language of this manuscript.

Authors' contributions

RK is the principal investigator that generated the study idea, is an operating surgeon, and revised and edited the manuscript. EB is the pediatric physician that followed the patients clinically, collected data, and revised the manuscript. AA wrote the introduction. HM wrote the case presentation, abstract, and discussion sections; collected the images; and reviewed the references. RA wrote the discussion and conclusion sections. All authors read and approved the final manuscript.

Funding

None received.

Availability of data and materials

All data generated or analyzed during this study are included in this published article.

Declarations**Ethics approval and consent to participate**

The study has been approved by Institutional Review Board (IRB). The IRB approval was granted through the University of Jordan review board on the 13th of January 2022.

Consent for publication

Written informed consent was obtained from the patients' parents for publication of this case report and accompanying images.

Competing interests

The authors declare they have no competing interests.

Author details

¹Pediatric Surgery Department, Jordan University Hospital, Amman, Jordan.

²Pediatric Department, Neonatal – Perinatal Division, Faculty of Medicine, Jordan University Hospital, University of Jordan, Amman, Jordan. ³Jordan University Hospital, Amman, Jordan.

Received: 11 March 2022 Accepted: 15 June 2022

Published online: 04 August 2022

References

1. El-Gohary Y, Gittes GK, Tovar JA. Congenital anomalies of the esophagus. *Semin Pediatr Surg.* 2010;19(3):186–93. <https://doi.org/10.1053/j.sempe dsurg.2010.03.009>.
2. Pinheiro PFM, Simões e Silva AC, Pereira RM. Current knowledge on esophageal atresia. *World J Gastroenterol.* 2012;18(28):3662–72. <https://doi.org/10.3748/wjg.v18.i28.3662>.
3. van Lennep M, Singendonk MMJ, Dall'Oglio L, et al. Oesophageal atresia. *Nat Rev Dis Prim.* 2019;5(1):26. <https://doi.org/10.1038/s41572-019-0077-0>.
4. Garge S, Rao KLN, Bawa M. The role of preoperative CT scan in patients with tracheoesophageal fistula: a review. *J Pediatr Surg.* 2013;48(9):1966–71. <https://doi.org/10.1016/j.jpedsurg.2013.06.010>.
5. Scot DA. Esophageal Atresia / Tracheoesophageal Fistula Overview. In: Adam MP, Ardinger HH, Pagon RA, et al., eds. 1993.
6. Teague WJ, Karpelowsky J. Surgical management of oesophageal atresia. *Pediatr Respir Rev.* 2016;19:10–5. <https://doi.org/10.1016/j.prrv.2016.04.003>.
7. Agha RA, Franchi T, Sohrabi C, et al. The SCARE 2020 Guideline: Updating Consensus Surgical CAse REport (SCARE) Guidelines. *Int J Surg.* 2020;84:226–30. <https://doi.org/10.1016/j.ijsu.2020.10.034>.
8. Prasad TR, Bajpai M. Intestinal atresia. *Indian J Pediatr.* 2000;67(9):671–8. <https://doi.org/10.1007/BF02762182>.
9. Farkash U, Lazar L, Erez I, Gutermacher M, Freud E. The distal pouch in esophageal atresia –to dissect or not to dissect, that is the question. *Eur J Pediatr Surg Off J Austrian Assoc Pediatr Surg=Zeitschrift fur Kinderchirurgie.* 2002;12(1):19–23. <https://doi.org/10.1055/s-2002-25091>.
10. Chen S, Chen D, Zhong W, et al. Saline-aided ultrasound versus upper gastrointestinal series in neonates and infants with suspected upper

gastrointestinal obstruction: a prospective multicenter comparative study. *AJR Am J Roentgenol.* 2022;218(3):526–33. <https://doi.org/10.2214/AJR.21.26621>.

11. Mahalik SK, Sodhi KS, Narasimhan KL, Rao KLN. Role of preoperative 3D CT reconstruction for evaluation of patients with esophageal atresia and tracheoesophageal fistula. *Pediatr Surg Int.* 2012;28(10):961–6. <https://doi.org/10.1007/s00383-012-3111-9>.
12. Basuguy E, Okur MH, Arslan S, Zeytun H, Aydoğdu B. A different approach to leakage of esophageal atresia in children. *Ulus travma ve acil cerrahi Derg Turkish J Trauma Emerg Surg.* 2020;26(6):927–31. <https://doi.org/10.14744/tjtes.2020.17745>.
13. Kulshrestha S, Kulshrestha M, Tewari V, et al. Conservative Management of major anastomotic leaks occurring after primary repair in esophageal atresia with fistula: role of extrapleural approach. *J Indian Assoc Pediatr Surg.* 2020;25(3):155–62. https://doi.org/10.4103/jiaps.JIAPS_73_19.
14. Godoy J, Ferraris T, Guelfand M. Early management of esophageal leak in esophageal atresia: changing paradigms. *J Laparoendosc Adv Surg Tech A.* 2020;30(8):923–6. <https://doi.org/10.1089/lap.2020.0099>.
15. Zhao J, Yang S, Li S, et al. Retrospective analysis of pneumothorax after repair of esophageal atresia/tracheoesophageal fistula. *BMC Pediatr.* 2021;21(1):543. <https://doi.org/10.1186/s12887-021-02948-x>.
16. Golden J, Demeter NE, Lim JC, Ford HR, Upperman JS, Gayer CP. Routine post-operative esophagram is not necessary after repair of esophageal atresia. *Am J Surg.* 2017;213(4):640–4. <https://doi.org/10.1016/j.amjsurg.2016.12.020>.
17. Baird R, Laberge J-M, Lévesque D. Anastomotic stricture after esophageal atresia repair: a critical review of recent literature. *Eur J Pediatr Surg Off J Austrian Assoc Pediatr Surg Zeitschrift fur Kinderchirurgie.* 2013;23(3):204–13. <https://doi.org/10.1055/s-0033-1347917>.
18. Sodhi KS, Saxena AK, Ahuja CK, Rao K, Menon P, Kandelwal N. Postoperative appearances of esophageal atresia repair: retrospective study of 210 patients with review of literature - what the radiologist should know. *Acta Radiol.* 2013;54(2):221–5. <https://doi.org/10.1258/ar.2012.120274>.

Publisher's Note

Springer Nature remains neutral with regard to jurisdictional claims in published maps and institutional affiliations.

Submit your manuscript to a SpringerOpen[®] journal and benefit from:

- Convenient online submission
- Rigorous peer review
- Open access: articles freely available online
- High visibility within the field
- Retaining the copyright to your article

Submit your next manuscript at ► [springeropen.com](https://www.springeropen.com)