

## A NEWBORN WITH DOUBLE ATRESIA: ESOPHAGEAL WITH DISTAL TRACHEOESOPHAGEAL FISTULA FOLLOWED BY CONCOMITANT ANAL ATRESIA - A CASE REPORT

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### Abstract

Esophageal atresia (EA) represents the most frequent congenital anomaly of the upper gastrointestinal tract in newborns with estimated incidence of one in 2500-4500 newborns worldwide. We present a rare case report of a newborn with double atresia, esophageal with distal fistula followed by anal atresia. A one-day-old male newborn was admitted to the Department of Pediatric Surgery at the Clinical Hospital Acibadem Sistina, Skopje, with the chief medical complaints of difficulty breastfeeding, vomiting and respiratory distress. After the initial clinical examination, and based on the medical history and physical findings, RTG revealed esophageal atresia with a distal fistula, followed by the identification of anal atresia. An urgent indication for surgical treatment was established, and a three-stage surgical plan was devised. In the first stage, the fistula was resected, followed by T-T esophageal anastomosis and sigmoidostomy. In the second stage, a laparoscopic pull-through Soave procedure was performed. The third and final surgical intervention was closure of the sigmoidostomy. On the seventh postoperative day (the primary procedure), an esophagogram showed adequate passage of contrast agent into the distal portion of the esophagus and stomach, with no fistulas. Neurological check-up showed normal EMG conducting of the anal sphincter. Esophageal atresia represents a surgical pathology with an overall survival rate of 85%-95%. The prognosis is corresponding to the presence of associated malformation. It is essential to emphasize that a thorough understanding of the anatomy of congenital malformations is indispensable to accomplish a successful repair and favorable outcomes in newborns.

**Keywords:** *newborn, double atresia, anal atresia, surgical management, neonatal surgery*

## Introduction

The esophagus is a muscular tube that transports a food bolus from the pharynx to the stomach. Embryologically, it is derived from the endodermal germ layer, which forms the pharynx, esophagus, stomach, and the epithelial lining of the aerodigestive tract. The trachea and esophagus arise from the separation of a common foregut tube during early fetal development. Failure of separation or complete development of this common foregut tube can lead to tracheoesophageal fistula (TEF) and esophageal atresia (EA)<sup>[1]</sup>. Esophageal atresia (EA) with tracheoesophageal fistula (TEF) is among the most common congenital anomalies requiring surgical intervention in infancy<sup>[2]</sup>. Prenatally, patients with EA may present with polyhydramnios, mostly in the third trimester, which may be a diagnostic clue to EA. Additionally, approximately 50% of patients with TEF/EA will have associated congenital anomalies including VACTERL (vertebral defects, anal atresia, cardiac defects, TEF, renal anomalies, and limb abnormalities) or CHARGE (coloboma, heart defects, atresia choanae, growth retardation, genital abnormalities, and ear abnormalities) syndrome. Once the neonate is born, the most common symptoms of EA include excessive drooling, choking, and failure to pass a nasogastric tube. Furthermore, if there is an associated TEF, gaseous distension of the stomach occurs as air passes from the trachea through the distal esophageal fistula into the stomach<sup>[1]</sup>.

Anal atresia is a congenital anorectal malformation featuring an absence of a normal anal opening and may be associated with a wide spectrum of congenital defects, ranging from a membranous obstruction to complex cloacal malformations involving the genital or urinary tracts<sup>[3]</sup>. The simultaneous occurrence of both anomalies in the same neonate, however, is exceedingly rare and poses significant diagnostic, therapeutic, and prognostic surgical challenges.

## Case report

In our case, the patient was born to a 35-year-old G2P1 mother via emergency cesarean section at 39 weeks of gestation due to fetal distress, with moderately meconium-stained fluid. The mother had a good antenatal history with regular intake of folic acid, iron, and calcium supplements with no other known comorbidities during and prior to her pregnancy. No previous pregnancies with congenital malformations were recorded, and family history was



**Fig.1.** Babygram of the patient



**Fig. 2.** Invert-gram of the same patient

unremarkable. Her antenatal scans were all unremarkable except for the evidence of polyhydramnios during the eighth month of her pregnancy. The baby was born with a weight of 2.7 kg and an Apgar score of 7/10 and 8/10 at 1 and 5 minutes, respectively. Three hours after delivery, the newborn began regurgitating milk during feeding. He appeared to be in distress, with signs of tachypnea, nasal flaring, grunting, subcostal retraction, and frothiness at the mouth. Later on, he was referred to our department by a pediatrician-neonatologist for further examination. An X-ray with a red rubber catheter *in situ* confirmed the diagnosis of esophageal atresia with distal TEF type C (Gross classification) (Figure 1). Further screening with an invertogram revealed concomitant anal atresia (Figure 2).



**Fig. 3.** Esophagography with contrast A-P



**Fig. 4.** Esophagography with contrast

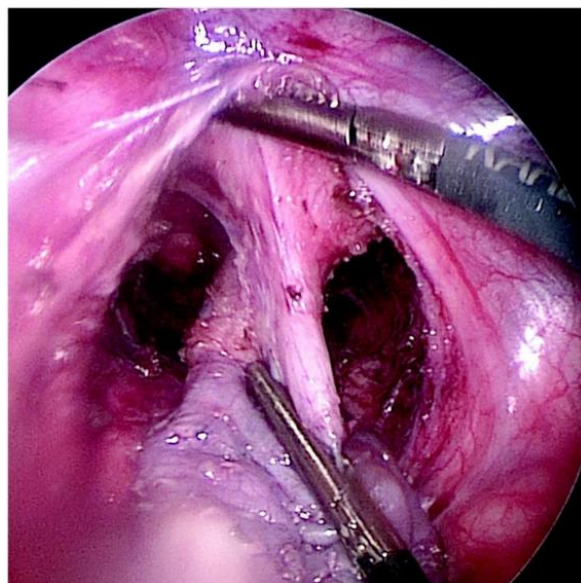


**Fig. 5.** Esophagography with contrast lateral position

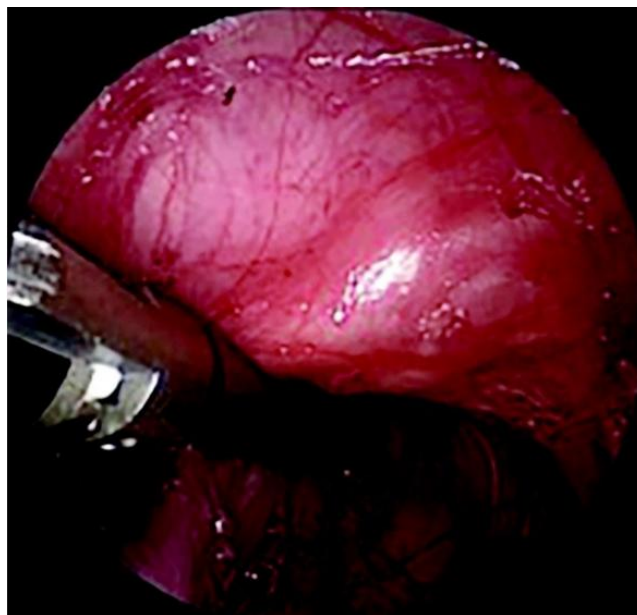


**Fig. 6.** Distal sigmoidography showing high ARM with rectovesical fistula

An urgent indication for surgical treatment including a three-stage surgical approach was established. After anesthesiologic evaluation and preoperative preparation, the baby was immediately transferred to the operating room. He was operated on via an open approach using an upper thoracotomy incision. In the first stage (primary, EA-repair), the esophageal fistula was resected, followed by creating a T-T esophageal anastomosis and a sigmoidostomy. On the seventh postoperative day, (Figures 3, 4 and 5), an esophagogram was performed, showing adequate passage of contrast agent into the distal portion of the esophagus and stomach, with no evidence of fistulas. Nine months after the initial esophageal reconstruction and sigmoidostomy, a preoperative control distal sigmoidography was performed (Figure 6), followed by laparoscopic-assisted pouch mobilization (LAARP) and rectovesical fistula ligation using a 5 mm stapler (Figures 7, 8, 9 and 10), representing staged ARM correction. The third and final surgical intervention was closure of the sigmoidostomy, following complex surgical management and ensuring the best possible outcome for the patient.



**Fig. 7.** Laparoscopy-assisted pouch mobilization



**Fig. 8.** Laparoscopy-assisted pouch mobilization and rectovesical fistula ligation using 5 mm stapler nine months after the initial esophageal reconstruction and sigmoidostomy



**Fig. 9.** LAARP – Laparoscopy-assisted pouch mobilization and rectovesical fistula ligation using 5 mm stapler



**Fig. 10.** ARM reconstruction following neurological EMG conducting of the anal sphincter

## Discussion

Esophageal atresia (EA) with or without tracheoesophageal fistula (TEF) is a congenital anomaly of the foregut characterized by discontinuity of the esophagus, often associated with an abnormal connection between the trachea and esophagus. It represents one of the most critical neonatal surgical emergencies and poses significant challenges in diagnosis, management, and postoperative care. The estimated global incidence of EA/TEF is approximately 1 in 2,500 to 4,500 live births, with no significant gender predilection, though slight male predominance has been noted in several studies<sup>[4]</sup>. Anal atresia, or imperforate anus, is another congenital malformation with an incidence of about 1 in 5,000 live births<sup>[5]</sup>. The simultaneous occurrence of both anomalies in the same neonate, however, is exceedingly rare and poses significant diagnostic, therapeutic, and prognostic challenges for pediatric surgeons. The coexistence of EA/TEF and anal atresia suggests a disturbance in embryological development during the critical period of foregut and hindgut differentiation. EA/TEF results from abnormal partitioning of the foregut into trachea and esophagus, while anal atresia arises from defective cloacal development and failure of the urorectal septum to descend properly.

When both anomalies coexist, the clinical presentation can be seriously complex: respiratory compromise may dominate the early clinical presentation, while abdominal distension and inability to evacuate stool become apparent later. This dual pathology can delay recognition of one anomaly when the other is more clinically urgent. Radiological evaluation plays a pivotal role. Failure of a nasogastric tube (NGT) to pass into the stomach suggests EA, while abdominal radiographs may reveal gas patterns consistent with distal fistula. Simultaneously, perineal inspection and invertogram or cross-table lateral radiographs confirm anal atresia. The combination of findings necessitates a multidisciplinary approach including neonatologists, pediatric surgeons, pediatric radiologists and anesthesiologists. The decision-making process must balance the urgency of correcting life-threatening EA/TEF against the need to establish fecal diversion for anal atresia. In most cases, EA/TEF repair is being prioritized due to the risk of aspiration and respiratory compromise; while colostomy for anal atresia can be performed either simultaneously or shortly thereafter.

Surgical repair remains the standard of care, with the primary aim of restoring esophageal continuity and separating the tracheal and esophageal lumina. Advances in neonatal anesthesia, preoperative stabilization, and postoperative intensive care have led to dramatic improvements in survival, with current success rates exceeding 90% in developed nations<sup>[6]</sup>.

*Survival and long-term outcomes depend on several factors:*

- **Associated anomalies:** The presence of cardiac, renal, or vertebral defects significantly influences prognosis.
- **Birth weight and prematurity:** Low birth weight and prematurity increase perioperative risk.
- **Timing and success of surgical interventions:** Early recognition and coordinated surgical management improve survival.

**Table 1.** Coexistence of esophageal atresia with distal TEF and anal atresia<sup>[7-9]</sup>

Author/Year	EA Type	ARM Type	Surgical Approach	Outcome
Spitz, 1996	EA + distal TEF	High ARM	Primary EA repair + staged ARM	Good survival
Levitt & Peña, 2010	EA + distal TEF	Low ARM	Primary EA repair + PSARP	Good continence
Jung, 2023	EA + distal TEF + duodenal atresia	High ARM	Thoracoscopic EA repair + staged ARM	Survival, mild GERD

Reports of double atresia are rare, with only isolated case series and anecdotal reports available, emphasizing the rarity of the condition and the importance of early diagnosis. Some

authors suggest that the coexistence of EA/TEF and anal atresia should prompt thorough evaluation for other anomalies within the VACTERL spectrum<sup>[2]</sup>. The rarity of such cases underscores the need for documentation, as each report contributes valuable insights into embryology, surgical strategy, and long-term outcomes (Table 1).

### Conclusion

Double atresia involving both the esophagus and anus is an exceptionally rare congenital anomaly. Our case highlights a newborn diagnosed with esophageal atresia (EA) with distal tracheoesophageal fistula (TEF), accompanied by anal atresia. The case underscores the diagnostic challenges, surgical management, and the importance of a multidisciplinary approach by an experienced team. Recent literature emphasizes innovations in minimally invasive repair, registry-based outcome studies, and the importance of long-term follow-up strategies. The modern surgical management of double atresia is particularly challenging due to the complexity of both anomalies demanding surgical correction, often in the neonatal period.

EA/TEF repair typically involves ligation of the fistula and primary esophageal anastomosis. Anal atresia repair depends on the level of the defect: low lesions may be corrected with perineal approaches, while high lesions often necessitate staged procedures with initial colostomy followed by definitive posterior sagittal anorectoplasty (PSARP)/LAARP – laparoscopic-assisted anorectoplasty. Early recognition, careful prioritization of surgical interventions, and multidisciplinary collaboration are essential to optimize outcomes. Given the rarity of this presentation, each case adds to the collective understanding of congenital malformations and informs future management strategies. Long-term follow-up is crucial for identifying potential complications and ensuring better quality of life for affected patients. Even with successful repair, patients may face long-term complications. EA/TEF survivors often experience gastroesophageal reflux, strictures, or dysmotility, while anal atresia patients may struggle with fecal continence and constipation. The coexistence of both anomalies compounds these challenges, necessitating prolonged follow-up and multidisciplinary care.

*Conflict of interest statement.* None declared.

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