

(CASE REPORT)



Association of esophageal atresia and duodenal atresia: A case report

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Abstract

Combined esophageal and duodenal atresia is a rare congenital malformation; its incidence is not well defined in the literature. Other associated malformations may include vertebral, anal, cardiac, renal and limb anomalies as part of a VACTERL syndrome.

We report an atypical case of association of esophageal and duodenal atresia in a newborn whose diagnosis of duodenal stenosis was made antenatally at ultrasound. and the diagnosis of esophageal atresia was made at birth in the delivery room.

The treatment of this malformation consists of an early surgical intervention to establish the digestive continuity. It requires ongoing multidisciplinary management to ensure proper growth and development of the newborn.

Keywords: Atresia; Esophageal; Duodenal; Association; VACTERL

1. Introduction

The association of esophageal atresia with duodenal atresia is extremely rare; it accounts for approximately 1-3% of all congenital esophageal malformations. The exact prevalence of this malformation is not well known, as it is extremely rare and varies among study populations [1].

The combination of these two congenital anomalies may occur sporadically or be associated with an underlying genetic syndrome, such as VACTERL syndrome (vertebrae, Anus, Heart, Trachea, Esophagus, Kidney, and Limbs) or CHARGE syndrome (Coloboma, cardiac anomalies, Nostril anomalies, Growth and/or developmental delay, Genital and/or Urinary anomalies, and Ear anomalies and/or hearing loss) [2-3].

This combination is often associated with significant morbidity and mortality. The management of this combination is controversial; it always leads to several management challenges, such as determining the order of treatment and the treatment regimen [2-4].

2. Observation

Our patient is a male neonate, preterm 37 SA, from a monitored pregnancy, non-consanguineous parents, mother 24 years old; negative infectious history; G2P1 with a previous neonatal death at D1 of life in a context of congenital heart disease, the delivery was by high route, Apgar 9/10/10, an antenatal ultrasound was done in favor of a strong suspicion of esophageal and duodenal atresia. The newborn was admitted for early respiratory distress.

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On admission, the clinical examination revealed facial dysmorphism, respiratory distress was rated 2-3/10 according to the Silverman score with polypnea and hypersialorrhea. The tube test was negative.

Thoracoabdominal radiography confirmed the diagnosis of esophageal atresia by showing gastric tube blockage at the upper esophageal cul de sac without gastric aeration. Echocardiography was normal. The transfontanellar (ETF) and renal ultrasound were also normal. The karyotype was without abnormalities.

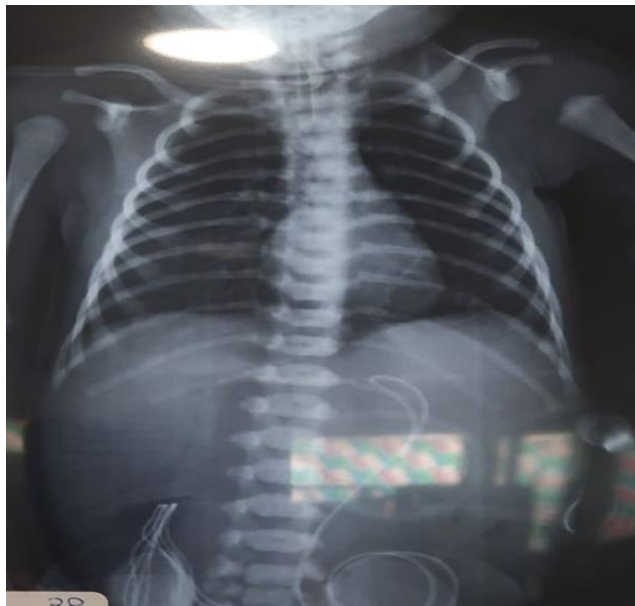


Figure 1 A thoraco-abdominal radiograph showing the gastric tube blocked at the level of the suprapubic area with a non-ventilated abdomen initially pointing to a type I or II esophageal atresia

The surgery was performed in 2 steps, a first exploration at D2 of life showing a complete duodenal atresia with the realization of a feeding gastrostomy. In a second step at D4 of life, by a thoracic approach, an atresia of the esophagus type I was confirmed and a cervical esophagostomy was performed.

The postoperative evolution was marked by a stay in a neonatal intensive care unit and the development of an inhalation pneumopathy in spite of being put on continuous soft suction and antibiotic therapy. The patient died at D21 of life in a refractory septic shock secondary to a sepsis with pulmonary origin.

3. Discussion

Esophageal atresia and duodenal atresia are relatively common congenital malformations, occurring in approximately 1 in 2,500 and 1 in 5,000 to 10,000 births, respectively [8]. The combination of these two anomalies is relatively rare and can present significant challenges to the health and nutrition of the newborn. Management will depend on the severity of each anomaly and their impact on the overall well-being of the patient. The protocol for management of this combination of anomalies is poorly defined and controversial. [6-7]

L.Spitz et al published a series of 18 neonates with esophageal atresia associated with duodenal atresia; 50 other associated malformations were identified in the 18 cases mainly genitourinary (11), cardiac (9), anorectal (8) and gastrointestinal (8) [3]. M. Miscia et al also presented in 2021 a cohort study of 74 newborns with esophageal atresia of which 5 cases had associated duodenal atresia which presents an incidence of 6.8% [1]. There was no consensus regarding the optimal treatment strategy for the case of association of both malformations [2]. Four of the five cases (80%) underwent simultaneous repair of both atresias, one of them with placement of a gastrostomy (25%). One out of five cases (20%) had a late diagnosis of duodenal atresia [1].

Surgical treatment should be performed as soon as possible, preferably within the first few days of life of the newborn, to minimize the risk of complications and optimize the chances of success. Most authors have recommended a staged repair of esophageal atresia combined with duodenal atresia with mandatory gastrostomy first, followed by duodenoduodenostomy a few days later [3-4]. Other teams have shown the effectiveness of simultaneous repair; there

was no apparent increase in morbidity and mortality with such an approach and they recommended the adoption of synchronous repair of both atresias in a single surgical procedure without the placement of a gastrostomy [1].

4. Conclusion

The combination of esophageal atresia and duodenal atresia is considered a rare and complex congenital malformation that requires early surgical intervention; multidisciplinary management to optimize long-term outcomes involving pediatric surgeons, neonatologists; nutritionists and speech-language pathologists.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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