Type IIIa Esophageal Atresia, Duodenal Atresia, and Intestinal Atresia in One of the Monochorionic, Diamniotic Twins: A Multi-Surgery Approach for Treatment

Cheng Tian, Jingsi Liu, Zhen Yang, Xianglun Sun, Ban Li, Kanglin Dai*
Department of Pediatric Surgery, The University of Hong Kong-Shenzhen Hospital, Shenzhen, Guangdong, China

Abstract: One of the monochorionic, diamniotic twins combined with type IIIa esophageal atresia, duodenal atresia, and intestinal atresia pose a rare management challenge. A female neonate from a monochorionic, diamniotic twin pregnancy who was diagnosed with multiple congenital malformations, including long-segment esophageal atresia with duodenal atresia, and intestinal atresia. These malformations were partially detected during prenatal ultrasound and were treated with a series of surgeries after birth. However, the patient continues to experience persistent esophageal stenosis, requiring regular dilation treatment. The treatment of multiple gastrointestinal malformations remains a significant challenge and may require multiple surgeries. Potential causes of defects and phenotypic differences in monozygotic twins, including genetic mutations, epigenetic factors, and non-genetic factors such as differences in amniotic sac size, uneven blood supply, and placental dysfunction. The probability of multiple digestive tract atresia occurring in one of the monozygotic twins is extremely low, and the reasons are diverse, requiring multiple surgeries.

***Keywords*:** Esophageal Atresia, Duodenal Atresia, Intestinal Atresia, Twin

1. Introduction

The live-birth prevalence of EA was 1.8 per 10,000 births[1]. Which is often associated with other anomalies such as vertebral anomalies, anal atresia, cardiovascular anomalies, tracheoesophageal fistula (TEF), renal anomalies, limb defects (VACTERL). The combined presence of long gap EA and DA and IA presents several management challenges. Interestingly, these abnormalities occurred in one of the monozygotic twins. should the defects be repaired together or in a staged way? This case presents our management strategy and patient outcome.

2. Patient

The patient is a female neonate naturally conceived and carried as a monochorionic, diamniotic twin by his 36-year-old mother. Standard prenatal care was followed. Fetal ultrasound at 22 week's gestation noted a linear cystic mass in the abdominal cavity of twin B, suggesting the possibility of intestinal duplication or jejunal atresia (Figure 1a). Besides, the weight of twin B was 15% lower than that of twin A. As the pregnancy progressed, the abdominal cyst persisted, in addition, the amniotic fluid volume of twin B increased. The patient was delivered by cesarean section at 31 weeks and 3 days due to premature rupture of membranes, with a birth weight of 1358 g and Apgar scores of 8/10.

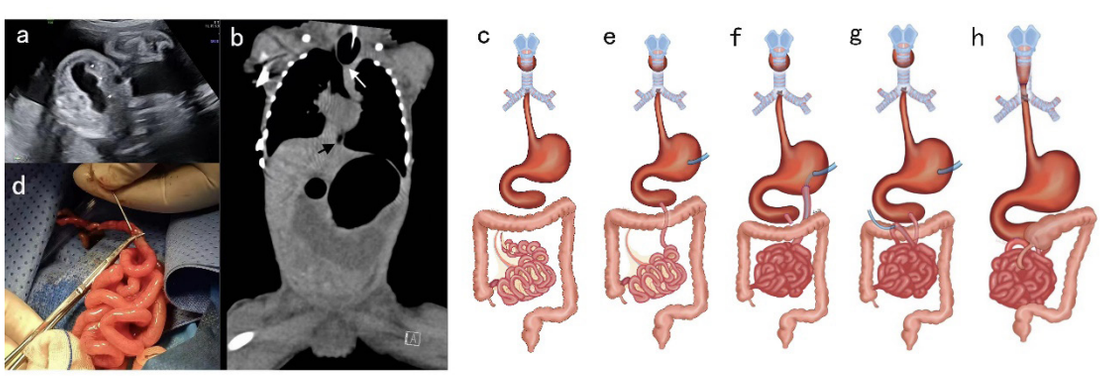

Figure 1. (a)Prenatal ultrasound indicates a linear cystic mass in the abdominal cavity; (b)Chest CT shows proximal esophageal atresia at approximately T2 (white arrow), distal esophagus visible (black arrow), and double bubble sign in the abdomen, indicating duodenal atresia; (c) Sketch of long segment esophageal atresia, duodenal atresia, and intestinal atresia of the patient; (d)The image of intestinal atresia;(e-h) Surgical sketch map.

Upon admission to the NICU, a radiography of the patient indicated a coil-up sign of the gastric tube in the esophagus and a distended gastric bubble, with an absence of distal intestinal gas. A CT scan confirmed long-segment esophageal atresia and tracheoesophageal fistula (T2-T4, type IIIa), coupled with dilated upper abdominal bowel (Figure 1b). Meanwhile, her sister's condition was stable.

On the third day post-birth, thoracoscopic TEF ligation and abdominal exploration were conducted, revealing duodenal atresia and two proximal jejunal atresia (Figure 1c). The severely affected small intestine was excised (Figure 1d), then the remaining 70 cm of small intestine was anastomosed to the duodenum. Subsequently, a gastrostomy was performed to facilitate postoperative enteral nutrition (Figure 1e).

The exploratory laparotomy was performed two months later due to digestive tract obstruction, notable adhesion between the duodenal-jejunal anastomosis and surrounding tissue was founded. The duodenal-jejunal anastomosis was disconnected, the duodenal stump was sutured, lastly the jejunum was anastomosed to the anterior wall of the stomach, allowing the gastrostomy tube to enter the jejunum (Figure 1f). However, one month following this procedure, the patient required another abdominal exploration due to gastrointestinal obstruction. Extensive intestinal adhesions were again identified. The gastro-jejunal anastomosis was severed, then the jejunum was re-anastomosed to the duodenum, finally a jejunostomy was performed (Figure 1g).

Thoracoscopic esophageal anastomosis was carried out four months after the birth. Subsequently, the patient developed a severe pulmonary infection, necessitating CT-guided pus puncture and catheterization.

One month later, the child exhibited symptoms of peritonitis. Abdominal radiography revealed free gas in the abdomen, prompting another exploratory laparotomy. Although no significant gastrointestinal perforation was detected, serious duodenal dilation indicated an obstruction at the duodeno-jejunal anastomosis. The jejunostomy was repositioned in the abdominal cavity and anastomosed with the duodenum. Unexpectedly, it was discovered that the ileocecal region was located in the upper left abdomen. Following the release of adhesions, the small intestine was relocated to the right abdomen, then the colon was positioned on the left side (Figure 1h). Afterwards, the child received regular esophageal dilation. In terms of prognosis, by the age of 2 years and 5 months, the patient measured 87 cm in height and weighed 11.5 kg. Her twin sister was 91 cm tall and weighed 13 kg. Both sisters demonstrated comparable intellectual and physical development. However, it is concerning that the patient continues to suffer from persistent esophageal stenosis, requiring regular dilation to date.

3. Disscusion

Midline structural defects are more common in monozygotic twinning[2]. Whole-genome sequencing analysis of 381 pairs of monozygotic twins showed that there were about 5.2 germline mutations between twins on average, which may lead to phenotypic inconsistencies in twins[3]. Besides, phenotypic differences in monozygotic twins are often not explained by genetic mutations alone and need to be combined with epigenetics[4] (eg, DNA methylation, X chromosome inactivation). Otherwise, differences in amniotic sac size, uneven blood supply, and placental dysfunction, can all lead to different phenotypes in monozygotic twins. To the best of our knowledge, a review of the literature suggests that this is the first case of a monochorionic-diamniotic twin that presented with duodenal atresia, esophageal atresia, and intestinal atresia, managed surgically. Given the potential for refractory stenosis resulting from anastomotic ischemia due to tension during the procedure, this patient may receive surgical resection and esophageal reconstruction in the future. Other treatment methods include endoscopic radical incision[5], local injection of triamcinolone acetonide and placement of absorbable stents[6], etc.

4. Conclusion

One of the monozygotic twins was diagnosed with long-gap EA, DA, and IA. The patient faced multiple surgeries and complications such as extensive postoperative adhesions. Currently, the infant's development and intelligence are normal, but there is persistent esophageal stenosis.

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