

## Triple Atresia: New Site, Same Fight!

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### Abstract

Triple atresia of the gastro-intestinal system usually comprises of atresia of the esophagus, duodenum and the anorectum. A 2-days-old neonate presented with frothing from mouth and absence of anal opening at birth. A red rubber catheter could not be passed into the stomach. Imaging showed the presence of a single large air bubble in the central abdomen. With the suspicion of the classical triple atresia; child was taken up for thoracotomy followed by laparotomy. The baby underwent fistula ligation and primary repair of esophageal atresia; pyloric web excision with Heineke- Mikulicz type pyloroplasty and a high sigmoid loop colostomy. The child succumbed on postoperative day 6 due to sepsis. This case of triple gut atresia left behind lessons about a new site of midgut atresia with the same fight for all to ponder upon and learn from.

**Keywords:** Triple Atresia; Esophageal Atresia; Tracheo-Esophageal Fistula; Pyloric Stenosis; Anorectal Malformation; Pyloroplasty

### Abbreviations

TA: Triple Atresia; EA+TEF: Esophageal Atresia with Trachea-Esophageal Fistula; DA: Duodenal Atresia; DS: Duodenal Stenosis; ARM: Anorectal Malformation; NG: Nasogastric; CPA: Congenital Pyloric Atresia

### Introduction

Triple atresia (TA) is an extremely rare occurrence of esophageal atresia (EA) with tracheoesophageal fistula (TEF), duodenal atresia (DA) or duodenal stenosis (DS), along with anorectal malformation (ARM) [1]. The combination of TEF with ARM is seen in about 4 - 9% cases [2,3]. However, the survival of such children despite surgery is gloomy, with a mortality of 24.1% [4]. Also, the combination of TEF with DA is rarer (1 - 2%) and the mortality is even higher, to the tune of 67% - 94% [5-8]. TA is even rarer, having the highest associated mortality [9]. Ours is a case of TA with an atypical site of the mid-gut atresia, pylorus, which has not been commonly reported earlier.

### Case Report

A two-days-old baby boy presented with frothing from mouth and an absence of anal opening since birth. Born to a primigravida at 36 weeks of gestation with no previous antenatal ultrasound available; the child was delivered at home. On presentation, he weighed 2010 gm and had a heart rate of 115/minute, respiratory rate of 60/minute with mild chest retractions, peripheral cyanosis, oxygen saturation of 89%, and delayed capillary refill time. On examination, a per-oral red rubber catheter could not be passed beyond 7 centimeters. The genitalia were male like with an absent anal opening. The perineum was flat with poorly developed gluteal folds. Spine and sacrum were normal. He had no radial limb defects.

After initial resuscitation, the X-Ray erect revealed coiling of the oro-gastric tube within the upper pouch with the presence of distal gas. A large gas shadow was observed in the central abdomen with paucity of gas in the pelvis (Figure 1). The routine blood investigations were normal except for a marginally decreased platelet count of 150,000. The echocardiography was suggestive of a small patent Ductus Arteriosus with left to right shunt, small atrial septal defect.

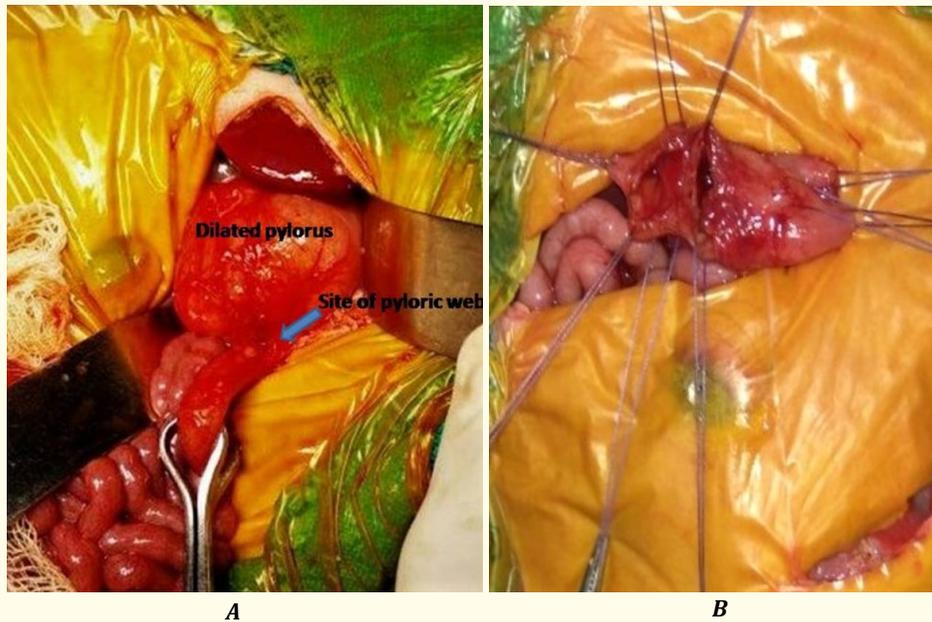


**Figure 1:** Babygram (erect) showing a large, central, gas shadow seen in the abdomen with the absence of gas distally in the pelvis; suggestive of mid-gut atresia.

A provisional diagnosis of esophageal atresia with tracheoesophageal fistula (EA+TEF) and associated anorectal malformation (ARM) was made; with a high suspicion of associated duodenal atresia. After adequate stabilization, the child was taken up for surgery.

The child was positioned in left lateral position and a standard right posterolateral thoracotomy incision made. The intra-operative findings were suggestive of a Type C-EA+TEF with a small fistula and a dilated, thick upper pouch with a gap of about one and a half vertebral bodies. The fistula was ligated and primary end-to-end esophageal anastomosis was performed under moderate tension over 5F nasogastric (NG) tube. Thereafter a laparotomy was performed via right upper transverse skin incision. A dilated enlarged stomach with a collapsed duodenojejunal and distal intestinal loops were observed. A suspicious region of thickened content was felt at the junction of the grossly enlarged pylorus and the first collapsed first part of the duodenum. A longitudinal incision was made along the antimesenteric border after taking stay sutures on either side. A complete pyloric web was identified; web excision with Heineke- Mikulicz type pyloroplasty was performed after ensuring distal patency (Figure 2). A high sigmoid loop colostomy was performed in view of high ARM and exteriorized through a separate incision. The surgical duration was 160 minutes and the blood loss was approximately 25ml. The baby was kept on elective ventilatory support thereafter.

The post-operative NG aspirate was bilious. From day 2, total parenteral nutrition was started. However he could not be weaned off his ventilatory support due to setting in of sepsis and later septicemic shock. Antibiotics were upgraded and inotropes were started, but he did not show much improvement. The X-ray Chest was suggestive of pneumonia on the 5<sup>th</sup> day. The child succumbed on postoperative day 6 due to pneumonia and septic shock.



**Figure 2:** Type 1 pyloric atresia with a complete diaphragm seen at the pylorus. A: Proximal pyloric end of stomach hugely distended and distal small intestinal loops seen as collapsed B: Enterotomy done longitudinally along the antimesenteric border of the pylorus and duodenum; complete diaphragm like web seen, excised and repair done transversely.

## Results and Discussion

Esophageal atresia with TEF is a common congenital anomaly. In 30% - 50% cases, it may be associated with other VACTERL (Vertebral, Anorectal, Cardiac, Tracheo-esophageal, Renal and Limb) anomalies [10]. The association of EA+TEF with DA has been reported in 1% - 2% of cases [2-5]. In such cases the diagnosis can be made clinically if there is failure to pass red rubber catheter beyond 7cm per orally in a neonate presenting with excessive drooling of saliva, and radiological picture is suggestive of double-bubble sign. A dye study through an abdominal esophagostomy/gastrostomy may sometimes be helpful in cases of pure EA who continue to have a high pre-feed gastric aspirate postoperatively, raising suspicion of a missed duodenal obstruction. Jackson., *et al.* [11] inferred from their study that the majority of the deaths in these patients of EA with DA were caused due to failure to recognize the second abnormality pre-operatively [11]. In addition to the above findings, an absent anal opening confirms the association of TA.

The usual cause of mid-gut obstruction in cases of TA is the DA or DS [1,9]. Our case is different from the usually reported cases of TA as the site of mid-gut obstruction was the pylorus. The most common anatomical type of congenital pyloric atresia (CPA) is type 1: pyloric membrane or diaphragm (60%), followed by type 2: pyloric atresia with no gap, and type 3: pyloric atresia with a gap [12]. The treatment depends upon the anatomical type. For Type 1 CPA, excision of diaphragm and Heineke- Mikulicz pyloroplasty has been described. The prognosis of isolated CPA has improved markedly over the years and is currently excellent. However, the overall mortality is more than 50% [13,14]. This is mainly because of the association of CPA with other fatal anomalies like Epidermolysis bullosa, aplasia cutis congenital, and multiple intestinal atresias [12].

The challenge in managing cases of TA remains the same; whether EA+TEF and ARM are associated with DA or CPA. Such babies usually undergo staged procedures [1]. However, with improved diagnostic modalities, anaesthesia techniques and mechanical ventilatory support; successful results in cases of antenatally diagnosed DA with a single staged repair of EA+TEF and DA have been described [8,15].

In our case also, a single staged repair of both EA+TEF and the pyloric stenosis seemed a feasible option. Al Salem, et al. [12] have reported a similar 2 days-old female baby, 2.4 kg, preterm who underwent fistula ligation, end to end esophageal anastomosis, gastro-duodenostomy and anal transposition who did well postoperatively and survived. In patients who are antenatally diagnosed (polyhydramnios, and double-bubble sign), institutional delivery, and post-natally well stabilized; a single staged repair of multiple intestinal anomalies is acceptable provided the surgical timing is kept in check.

The second controversy in TA patients undergoing staged repair is whether to repair the TEF first or to perform it later after a gastrostomy and colostomy have been done. In sick and unstable babies who present late or who present with perforation peritonitis, a colostomy and gastrostomy help tide over the initial crisis of abdominal distension, respiratory compromise and severe sepsis. Once stabilized, the baby can be taken up for a second surgery of TEF repair [1]. On the other hand, some patients who have undergone EA+TEF repair first and are diagnosed with DA post-operatively through dye study, have also been reported to do well after DA repair later on [1].

The previously reported morbidity and mortality in patients of TA has been high [9], though recent improvement has been observed [1,12]. The most common cause of death has been sepsis, like in our case.

### **Conclusion**

There is no doubt that TA is an extremely rare combination. However, due to scarcity of literature on such cases, no management protocol for these patients can be formulated or generalized. High index of suspicion, awareness about this condition, early diagnosis and a staged and timely repair are the key to its management. The site of mid-gut atresia may vary from pyloric stenosis, pyloric atresia, to duodenal stenosis or atresia and annular pancreas. The surgery consists of ligation of TEF and repair of EA, followed by duodenoduodenostomy/ web excision and pyloroplasty with colostomy. Parental nutrition is mandatory. Mortality is high and prognosis is poor if not managed in time. Pre-operative resuscitation and surgical expertise have a major role in improving survival in patients with TA.

### **Conflict of Interest**

Authors declare that no conflict of interest exists.

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