

Congenital Triple Atresia: A Diagnostic
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Abstract

One day preterm female neonate was presented with excessive drooling of saliva and non passage of meconium since birth to the Department of Pediatric with antenatal history of polyhydramnios detected at 28 weeks followed by fetal distress at 32 weeks which leads to preterm delivery. An X-ray of the chest and abdomen with a nasogastric tube showed coiling of nasogastric tube in the upper oesophageal pouch in the upper chest and a gasless abdomen confirming the diagnosis of pure oesophageal atresia; child had absent anal opening with rectovaginal fistula which was confirmed by passing small feeding tube in the fistula. Oesophagostomy and feeding gastrostomy were done at first surgery. On 2nd postoperative day child developed intolerance and blockage to feed after first few feeds. Contrast dye study done through feeding gastrostomy showed duodenal atresia which was missed during first surgery due to small, collapsed stomach and duodenum due to no gas in the abdomen due to pure oesophageal atresia. Second surgery duodenoduodenostomy was done on 3rd postoperative day of first surgery, but child succumbed postoperatively due to septic shock and prematurity.

Introduction

The association of esophageal atresia or EA with Tracheoesophageal Fistula (TEF) and Duodenal Atresia (DA) or Duodenal Stenosis (DS) is well-recognized, although uncommon [1,2]. The occurrence of triple atresia (TA), that is, EA with or without TEF, DA, or DS, along with Anorectal Malformation (ARM) is extremely rare, hence, managing such cases is extremely difficult for the treating pediatric surgeons [1,2].

The combination of ARM along with Oesophageal Atresia (EA) with or without Tracheoesophageal Fistula (TEF) is seen in about 4-9% cases [3,4].

The management of triple atresia is challenging. These patients are usually managed by staged procedures [3-5].

Conventionally, several studies have demonstrated that oesophageal atresia should be repaired primarily, followed by other surgeries [5,6].

In contrast, other investigators have demonstrated that oesophageal atresia repair should be done as the second surgery after colostomy and gastrostomy [7-10].

Combination of oesophageal atresia with DA is rarer and the mortality is even higher, to the tune of 67% - 94%. Due to associated congenital abnormality [3,4,6,7].

Case Report

One day preterm female neonate presented with excessive drooling of saliva and non passage of meconium since birth to the Department of Pediatrics in our institute. There was antenatal history of polyhydramnios detected at 28 weeks followed by fetal distress at 32 weeks of pregnancy for which emergency Caesarean section delivery was done. Birth weight was 1.6 kg.

It was on examination not possible to pass nasogastric tube. The nasogastric tube got coiled in the mouth and there was absent anal opening with high rectovaginal fistula identified (Anorectal malformation). An X ray of the chest and abdomen with a nasogastric tube was done, which showed coiling of nasogastric tube in the upper oesophageal pouch in the upper chest and a gasless abdomen confirming the diagnosis of pure oesophageal atresia; (Figure 1). Pediatric Surgeon opinion was taken and after adequate stabilization, the child was taken up for the surgery on day 2 of life. Intraoperatively upper blind pouch was brought as oesophagostomy (Figure 2) and on opening abdomen stomach was small, collapsed, duodenum and whole small bowel were collapsed due to absent gas in the abdomen due to pure oesophageal atresia, feeding gastrostomy was done. As child had absent anal opening with rectovaginal fistula which was confirmed by passing a small feeding tube in fistula (Figure 3), hence colostomy was abandoned.

On 2nd postoperative day tried to give feeds through gastrostomy tube, but there was intolerance and blockage to feeds after first few feeds. Hence contrast dye study was done through feeding



Figure 1: Showing coiling of nasogastric tube in the upper oesophageal pouch in upper chest and a gasless abdomen suggesting pure oesophageal atresia.



Figure 4: Showing contrast study through feeding gastrostomy; no contrast seen beyond duodenum after 6 hours of serial X-ray abdomen and there was no gas in the abdomen suggest duodenal atresia.



Figure 2: Upper blind pouch was brought as oesophagostomy with feeding tube in situ.



Figure 3: Shows recto-vaginal fistula with absent anal opening confirmed by passing small feeding tube in fistula.

gastrostomy tube and found no contrast going beyond duodenum after 6 hours of serial X-ray abdomen and also there was no gas in the abdomen hence diagnosed to have duodenal atresia (Figure 4). Haematological investigation showed raised CRP, platelets was 20000, sepsis was suspected, blood culture was sent, and antibiotic upgraded, Pediatric surgeon consultation was taken and planned for 2nd surgery for duodenal atresia after platelet transfusion and stabilization on next day.

During second surgery duodenal atresia of second part of the duodenum was found, which was missed during first surgery due to gasless abdomen due to pure oesophageal atresia. Duodenoduodenostomy was done and child shifted to NICU.

Postoperatively inotropes were started considering hemodynamic stability. Child started deteriorating on 2nd day of second surgery; baby was kept on ventilatory support thereafter. However baby could not be weaned off from ventilator support due to setting in of sepsis and later septicaemic shock. Antibiotics were upgraded and inotropes were added on and dose was increased, but baby did not show much improvement. The child succumbed on postoperative day 4 of second surgery.

Discussion

On antenatal USG, oesophageal atresia is usually suspected when there is polyhydramnios and an absent or small stomach [11]. This, however, is not definitive; the positive predictive value of these findings is only 56% [12]. In our case there was antenatal history of polyhydramnios which was detected at 28 weeks followed by fetal distress at 32 weeks which leads to preterm delivery.

Theoretically, a diagnosis of EA with or without TEF, DA, and ARM can be easily done by the plain radiographic study of the chest and abdomen, but practically chances of missing DA are very high when they are associated with pure EA. Quite often, the diagnosis is

suspected when gastrostomy or a nasogastric feed are not tolerated by the child in the early postoperative period following thoracotomy. A high index of suspicion of atresia should be kept in mind before considering gastroesophageal reflux as a possible cause of high prefeed aspirate [13]. In our case also we missed duodenal atresia during first surgery due to small, collapsed stomach and duodenum due to absent gas in the abdomen due to presence of pure oesophageal atresia and duodenal atresia was diagnosed by contrast dye study through feeding gastrostomy on 2nd postoperative day when child developed intolerance and blockage to feed after first few feeds.

Spitz [6] in a review of 18 patients of oesophageal atresia and duodenal atresia emphasized that these babies are at high risk with mortality rates ranging from 67% to 94% [6]. Jackson and colleagues [14] infer that the majority of these deaths are caused by failure to recognize the second abnormality pre-operatively [14], in our case also child succumbed postoperatively due to septic shock and prematurity.

High index of suspicion, awareness about this condition, early diagnosis, staged and timely repair are the key to its management. Mortality is high and prognosis is poor if not managed in time. The most common cause of death has been sepsis [15], like in our case.

Conclusion

Awareness of Triple atresia, High index of suspicion in every case of Oesophageal atresia or Anorectal malformation, early diagnosis and timely repair are the key to reduce mortality and improve prognosis.

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