

Abnormal Presentation of Type III Oesophageal Atresia and TOF

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Introduction:

Tracheoesophageal fistula (TOF) is a congenital communication between the trachea and oesophagus. TOF often leads to severe and fatal pulmonary complications. It is usually combined with esophageal atresia. More than 95 types had been described in the literature up to 1976⁽¹⁾. Since then newer subtypes also were described⁽²⁻⁷⁾. The commonest one is type iii. The latter is diagnosed by suggestive clinical presentation and gaseous abdomen in the X-Ray. Gasless abdomen in this type of oesophageal atresia is an extremely rare condition and makes a diagnostic dilemma.

Case Report:

A male neonate of seven days from rural Gezira was referred to our center with shortness of breath and excessive salivation since birth. Pregnancy was complicated with polyhydramnios which was discovered during the antenatal period by ultrasound.

The baby was delivered at home vaginally, cried immediately, but was noticed to have cyanosis and choking during breastfeeding. So breast feeding was withheld. The baby passed meconium in the first day. He was looking unwell, but not cyanosed nor dehydrated. The pulse rate was 162 bpm and respiratory rate is 60 cpm. There were active alae-nasi and intercostals recession. The abdomen was scaphoid.

During the insertion of a radio-opaque nasogastric tube, the tube's insertion stopped in the oesophagus about 8 cm from its advancement in the nostril. Chest X-ray showed coiled nasogastric tube in the upper chest (Figure 1). Abdominal XRay showed gasless abdomen (Figure 2). On this clinicoradiological background the diagnosis of pure oesophagel atresia (type I) was made. Other anomalies were excluded. The baby underwent oesophagostomy and gastrostomy as a first stage. On

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the second postoperative day the baby was fed through the gastrostomy tube, but the milk was noticed coming out through the esophagectomy. The baby underwent definitive surgery four days later. The intra-operative findings were an esophageal atresia (fibrous band in between the 2 ends of the oesophagus) and a distal tracheoesophageal fistula. The fistula was found closed by a mucus plug. Postoperative course passed uneventful. Two months later the baby developed esophago-cutaneous fistula which was repaired without complications.



Figure 1: Chest x-ray shows Coiled NG tube appearing in the upper chest



Figure 2: Erect abdominal X-Ray shows no gastric bubble

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Discussion:

EA is a condition in which the proximal and distal portions of the esophagus do not communicate. The upper segment of the esophagus is a dilated blind-ending pouch with a hypertrophied muscular wall. The pouch typically extends to the level of the second to fourth thoracic vertebra. In contrast, the distal esophageal portion is an atretic pouch with a small diameter and a thin muscular wall; it usually extends 1-2 cm above the diaphragm. When it is associated with TOF, the fistula commonly enters the trachea posteriorly just above the carina. However, isolated TOF, or an H-fistula, can occur at any level from the cricoid cartilage to the carina. The table below describes the 5 main categories of congenital TOFs (Table 1).

In our case, it was the type III OA/TOF, which was classically diagnosed by clinical presentation and gaseous bowel distention on the X-ray. The latter finding was not found and this leads us to the diagnosis of pure OA preoperatively. Passage of milk through the oesophagostomy alerted us to revise the initial provisional diagnosis. The explanation of this was discovered intraoperatively, were a fistula occluding mucous plug was found and this prevented the gas from entering the stomach.

Table 1: Classification of Congenital Tracheoesophageal Fistulas and Esophageal Atresia

Anatomic Characteristics	Percent of Cases
Esophageal atresia with distal TOF	87
Isolated esophageal atresia without TOF	8
Isolated TOF	4
Esophageal atresia with proximal TOF	1
Esophageal atresia with proximal and distal TOF	1

Occlusion of the lower segment by a mucus plug has been described by other authors ⁽⁸⁾. Other possible reasons for a gasless abdomen can be: (1) one or more atresias in the lower segment ⁽⁹⁾; or (2) a high degree of stenosis in the lower segment. ⁽⁸⁾

Our conclusion and recommendation that paediatric surgeon should consider the anatomical variations and their effects on clinical and radiological findings before surgical treatment.

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