Review article on

Tracheoesophageal Fistula

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1 Introduction

The trachea or the windpipe carries air to the lungs whereas the esophagus carries food to the stomach. A tracheoesophageal fistula (TEF) is an abnormal connection between these two tubes. It is a common congenital abnormality .When this happens, air within the trachea may pass into the esophagus or alternatively, food passes through the trachea.

Tracheoesophageal (TE) fistula is a common congenital anomaly that occurs in 1 out of every 3,500 live births. TE fistula is commonly associated with other congenital malformations, particularly cardiac defects. Esophageal atresia is a related congenital malformation with a presentation similar to that of a TE fistula and can occur with or without the presence of a fistula.

Definition:

Esophageal atresia is caused by a failure of the esophagus to develop as a continuous passage from the throat to the stomach resulting in obstruction of the infant's normal swallowing route.-

A/c Marlow

Congenital atresia of the esophagus and tracheoesophageal fistula (TEF) are rare malformations that result from failed separation of the esophagus and trachea by the fourth week of gestation.

- A/c WONG'S

Esophageal atresia refers to a congenitally interrupted esophagus,where the proximal and distal ends do not communicate the upper esophageal segment ends in a blind pouch and the lower segment ends a variable distance above the diaphragm

- A/c Lippincott

Incidence:

The incidence of TEF is approximately 1 in 3500 births. EA and TEF are classified according to their anatomic configuration. Type C, which consists of a proximal esophageal pouch and a distal TEF, accounts for 84% of cases. TEF occurs without EA (H-type fistula) in only 4%.

Etiology:

EA/TEF occurs as a result of a developmental failure during early fetal (embryonic) growth. The reason this failure occurs Is not fully understood. Isolated EA/TEF is believed to occur due to the combination of multiple different factors (multifactorial inheritance). Such factors can potentially include genetic, environmental, and immunological ones. However, no specific contributing factors have been identified. Most cases of isolated EA/TEF occur sporadically and the risk of recurrence in subsequent pregnancies is less than 1 percent. Tracheoesophageal fistula occurs in 1 in 3500 births, with slight male dominance. Esophageal atresia with or without tracheoesophageal fistula is common in prematurity, with 34% of cases weighing less than 2500 grams.

Approximately 50%of neonates with esophageal atresia or tracheoesophageal fistula have other anomalies also that are represented by acronym "VACTERL".

- **VACTERAL Syndrome its combination of**
  - ✓ V - Vertebral column defects
  - ✓ A - Anorectal malformation
  - ✓ C - Cardiovascular
  - ✓ T - Tracheoesophageal fistula
  - ✓ R - Renal anomalies
  - ✓ L - Limb Anomalies
- Low weight of the baby
- Prematurity
- History of polyhydramnios
- Infants with defects.

TYPES:

- **Type A:** There's no TEF, only EA. Esophagus is divided into two parts, with both portions ending in blind pouches (cavities that are closed at one end). This is also commonly referred to as pure esophageal atresia. It makes up about 8% of all cases.
- **Type B:** This rare form of TEF affects about 2% of all cases. The lower portion of esophagus ends in a blind pouch, and the upper portion are connected to windpipe by a tracheoesophageal fistula.
- **Type C:** The most common form of TEF, type C is when the upper portion of esophagus ends in a blind pouch, and the lower portion is connected to trachea by a tracheoesophageal fistula. About 85% of babies born with this type.
- **Type D:** In this rarest form of TEF, a tracheoesophageal fistula connects both the upper and lower portions of esophagus to trachea. Less than 1% of babies born with this type.

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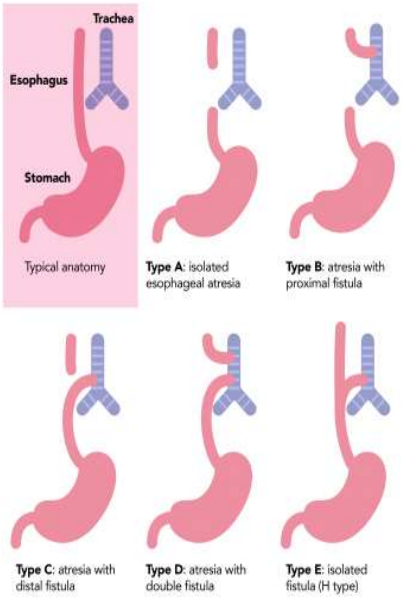
❖ **Type E:** Esophagus connects to stomach normally and is fully intact. However, a tracheoesophageal fistula connects esophagus and trachea. Type E affects 4% of TEF cases.



2 Pathophysiology

The Upper part of esophagus is developed from the retropharyngeal segment and lower part from the pregastric segment of the foregut. At 4 weeks of gestation a laryngeal groove is formed which divides the foregut in two longitudinal tubes, which further develop in to the respiratory tract and digestive tract. Defective separation due to deviated or incomplete septum (or) incomplete fusion of tracheal folds results in malformation of trachea and esophagus.

3 Clinical manifestations

Affected infants may experience episodes of coughing, gagging or choking. They can also experience repeated episodes of low levels of oxygen in the blood (hypoxia/cyanosis). Cyanosis is characterized by shortness of breath, coughing, flaring of the nostrils when breathing and bluish discoloration of the skin. Some of these infants have abnormal softening and weakening of the cartilage of the trachea (tracheomalacia) so that the walls of the trachea are floppy instead of rigid. This is often mild, but can be severe, leading to collapse of the air passage. Tracheomalacia can contribute to breathing difficulties and may precipitate respiratory arrest (near death episodes).

4 Diagnostic Evaluation

- A Clinical evaluation for any potentially associated congenital defects, especially heart defects, will collecta through health history and assess the feeding difficulties.
- The diagnosis of EA/TEF is confirmed by clinical testing i.e., attempting to pass a nasogastric tube down the throat of infants who have require excessive suction of mucus, or are born to mothers with polyhydramnios, or, if earlier signs are missed have difficulty feeding.
- A routine ultrasound during pregnancy. But most TEFs are diagnosed soon after birth, based on an infant's symptoms.
- X-rays of the chest and stomach, and
- Esophageal endoscopy or bronchoscopy.
- Echocardiogram, to rule out cardiac irregularities.

5 Treatment:

Treatment may require the coordinated efforts of a team of specialists. Pediatric surgeon, paediatrician, cardiologists, speech therapist, intensive care specialists and nurses, and other healthcare professionals may need to systematically and comprehensively plan a program for the child's treatment and ongoing care. Treatment is best performed at tertiary referral hospitals that are well versed in treating these conditions.

A supportive care to prevent aspiration is necessary. A suction catheter is placed in the pouch at the end of the upper portion of the oesophagus. A catheter is a hollow flexible tube. The suction catheter continually sucks out secretions that accumulate in the pouch to prevent aspiration. Broad spectrum antibiotics may be given to the infant if sepsis or pulmonary infection is present or suspected. In infants with respiratory failure, additional measures such as assisted ventilation may be required but should be used with extreme caution.

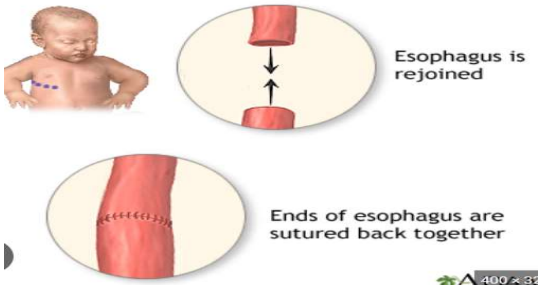
6 Management

Esophageal atresia is surgical emergency,the treatment includes maintenance of a patent airway, Prevention of pneumonia and gastric or blind pouch decompression and surgically repair of the anomaly.

Therapeutic interventions:

- Infant is immediately taken off oral intake
- Infusion of I.V fluids.
- Maintain the position(supine or upright position) to control the aspiration
- Removal of secretions from the mouth and upper pouch requires frequent or continuous suction
- Because aspiration pneumonia is almost inevitable and early broad spectrum Antibiotic therapy

A Thoracotomy: It is a primary surgical correction division and ligation of TEF and an end -to – side anastomosis of the esophagus.This may consist of one operation or be staged with two or more proceduresFor Infants who are preterm have multiple anomalies or in poor condition a staged procedure that involves palliative measures including Gastrostomy, ligation of the TEF and provision of constant drainage of the esophageal pouch. A delayed esophageal anastomosisis usually attempted after several weeks to months when the upper pouch elongates. Further surgical techniques may be performed later to facilitate esophageal lengthing. If any esophageal anastomosis still cannot be a ccomplished,Cervicalesophagostomy (to allow drainage of saliva) and Gastrostomy are performed



Complications include:

- A leak of saliva where the two ends of the esophagus were connected
- Gastroesophageal reflux (GERD) is very common after EA/TEF surgery.
- The newly connected esophagus may not function perfectly at first.

7 Prognosis

The prognosis is unsatisfactory and depends upon early diagnosis whether pneumonia has occurred or notpresence of serious anomalies such as congenital heart diseasesStandard medical and nursing care.

8 Nursing management:

Assessment of an infant with tracheoesophageal atresia includes:

- **History andPhysical exam.** The acronym VACTERL (vertebral defects, anorectal malformations, cardiovascular defects, tracheoesophageal defects, renal anomalies, and limb deformities) refers a set of associated anomalies that should be readily apparent upon physical examination

9 Nursing Diagnosis

- Impaired gaseous exchange related to abnormal opening between oesophagus and trachea as evidenced by cyanosis.
- Impaired swallowing related to mechanical obstruction.
- Risk for injury related to surgical procedure.
- Anxiety related to difficulty swallowing, discomfort due to surgery.
- Altered family processes related to children with physical defects.
- Risk for aspiration related to difficulty in swallowing.

10 Nursing Interventions:

- **Ensure safe swallowing.** Place suction equipment at the bedside, and suction as needed; ensure proper nutrition by consulting with physician for enteral feedings.
- **Prevent aspiration.** Check placement before feeding, using tube markings, x-ray study (most accurate), pH of gastric fluid, and colour of aspirate as guides; if ordered by physician, put several drops of blue or green food colouring in tube feeding to help indicate aspiration. In addition, test the glucose in tracheobronchial secretions to detect aspiration of enteral feedings; elevate the head of bed to 30 to 45 degrees while feeding the patient and for 30 to 45 minutes afterward if feeding is intermittent; and instruct in signs and symptoms of aspiration.

Reduce anxiety. Allow family caregivers to talk about anxious feelings and examine anxiety-provoking situations if they are identifiable; assist them in developing new anxiety-reducing skills (e.g., relaxation, deep breathing, positive visualization, and reassuring self-statements); explain all activities, procedures, and issues that involve the patient; use nonmedical terms and calm, slow speech; do this in advance of procedures when possible, and validate patient's understanding.

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