Unintentional Esophageal Intubation in a Newborn with a Tracheoesophageal Fistula with Unknown Tracheal Atresia: A Case Report

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Case Description

Our patient is a 2-day old, 1.64kg, female born at 33 weeks gestation, who was transferred to Rush University Medical Center (RUMC) with respiratory distress with presumed TEF and esophageal atresia after difficulty placing NG tube. Upon arrival to RUMC, the patient was stable, with endotracheal tube in place. The patient was taken to the OR for TEF repair. The patient was placed on pressure support, with appropriate monitoring in place. End-tidal CO2 monitoring was inconsistent from the start, thought to be secondary to an audible air leak. Despite inconsistent EtCO2, other vital signs were stable, so the decision was made to keep the ETT in place.

Anesthetic Course

In the OR, ligation of the distal esophagus from the trachea was performed. Per surgeon request, a bougie was placed to identify the proximal esophageal pouch. Upon insertion, EtCO2 was lost, and the patient began to desaturate. We were unable to manually ventilate the patient via the ETT. The decision was made to remove the ETT. Bag-mask was performed without significant difficulty, with improvement of SpO2.

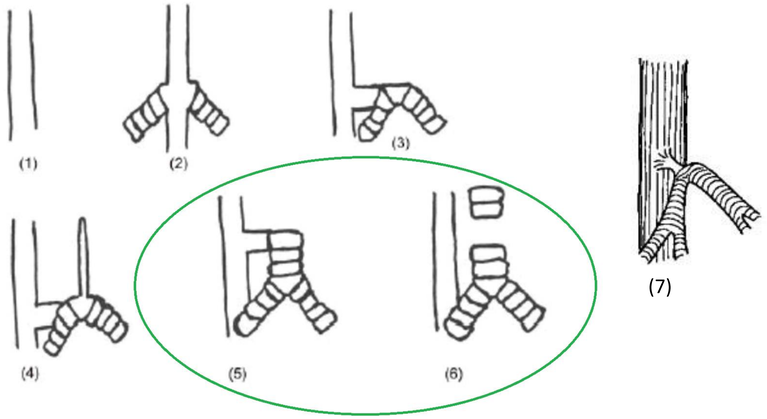
Endotracheal intubation was attempted in the lateral position, as the chest was open, however unsuccessful via direct laryngoscopy and fiberoptic scope. We were able to bag-mask effectively in between. The chest wall was closed, and the patient was placed in the supine position. An additional pediatric anesthesiologist was called into the OR for assistance. Numerous attempts at endotracheal intubation were made via direct laryngoscopy and fiberoptic scope, however we were unable to obtain EtCO2 and the patient began to desaturate, despite a Grade I view.

The decision was made to contact the Otolaryngology (ENT) service. Rigid bronchoscopy was performed, and it was quickly apparent that the proximal and distal trachea were in discontinuity at the subglottic level, with the proximal trachea opening directly into the mediastinum.

The chest was reopened, and the trachea was accessed distally, just above the carina. A 2.0mm cuffless ETT was passed without difficulty, and connected to the anesthesia circuit with positive EtCO2. ENT then passed a 2.0mm cuffless ETT through the oral cavity, into the esophageal remnant. The ETT was located by the pediatric surgeons in the chest, and directed into the airway through an additional TEF. The patient was then taken to the NICU.

Tracheal Atresia/Agenesis

Tracheal agenesis is a rare, and often fatal, congenital anomaly that results in complete interruption or absence of the trachea. It occurs in less than 1/50,000 births (2), with only a few published cases worldwide. Of the reported cases, most are associated with co-existing congenital abnormalities, including cardiac, pulmonary, gastrointestinal, genitourinary, musculoskeletal, and CNS defects. Without a co-existing tracheoesophageal fistula (TEF), these patients are unable to survive. When a TEF is present, survival is rare, yet possible with timely diagnosis, careful planning and surgical correction. Ventilation must be achieved via bag-mask, or endotracheal tube through the esophagus. Long-term survival is dependent on maintaining a stable airway, and correcting any concomitant anomalies. Based on a literature search, there have been only 5 cases of long-term survival (3).



(7)

Faro/Floyd Classification System (1)

1 – Faro A
2 – Faro B/Floyd III
3 – Faro C
4 – Faro D
5 – Faro E/Floyd I
6 – Faro G
7 – Floyd II

References:

1. Duarte, S., et al. (2016). When Only Esophageal Intubation Assures Ventilation: A Case Report. Journal of Anesthesia and Clinical Research, 7(11).

2. Ergun, S., et al. (2011). *Tracheal agenesis: A rare but fatal congenital anomaly.* McGill Journal of Medicine, 13(1), p. 10.

3. Fuchimoto, Y., et al. (2011). *A long-term survival case of tracheal agenesis: management for tracheoesophageal fistula and esophageal reconstruction.* Pediatric Surgery International, 27(1), p. 103-106.

Perioperative Management Goals in Patients with Tracheal Atresia/Agenesis

- Clinical suspicion (3)
  - Pre-natal U/S → limited use when TEF present
    - Polyhydramnios
    - Fetal Ascites
    - Dilated Trachea
    - Fluid in upper airways
    - Hyper-echogenic lungs
    - Fetal MRI
  - Absent cry at birth
  - Respiratory distress
  - Ability to mask ventilate
  - Unable to pass ETT despite direct view of vocal cords

- <u>Pre-operatively</u>
  - Inter-disciplinary discussion with OB/GYN, ENT, NICU, and pediatric anesthesiologist, if possible → plan
  - Proper equipment available
  - Bag-mask + esophageal intubation proximal to or into fistula (spontaneous ventilation)
  - Further imaging if desired

- <u>Intra-operatively</u>
  - Tracheostomy (if possible/required), esophageal reconstruction/ligation/esophagostomy, gastrostomy, airway reconstruction
  - Confirm ETT placement
  - EXIT procedure management, if planned

- <u>Post-operatively</u>
  - Discussion with the team regarding complications and extubation
  - Plan for re-intubation
  - Nutritional goals
  - Long-term goals

Resolution

The patient ultimately passed away in the NICU 8 days after her procedure secondary to respiratory failure