When Only Esophageal Intubation Assures Ventilation: A Case Report

Sonia Duarte 1, Jacinta SA 1, Pedro Pina 1 and Humberto S Machado 1,2*

1Anesthesiology Department, Centro Hospitalar do Porto, Porto, Portugal
2Instituto de Ciências Biomédicas Abel Salazar, Universidade do Porto, Porto, Portugal

Corresponding author: Humberto S Machado, Anesthesiology Department, Centro Hospitalar do Porto, Largo Abel Salazar, 4099-001 Porto, Portugal, Tel: 351 935848475; Fax: 351 220900644; E-mail: hjs.machado@gmail.com

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Abstract

Tracheal defects can lead to difficulties in airway management in newborns. Tracheal atresia is a very rare entity with high mortality that presents a challenge to the anesthesiologist. We report a case of a newborn with tracheal atresia, esophageal atresia and double tracheoesophageal fistula, which ventilation, until the 6th day of life, was assured through an esophageal intubation.

Keywords: Airway; Newborn; Trachea; Tracheoesophageal fistula; Tracheal stenosis; Tracheal agenesis

Introduction

Trachea and esophagus begin their independence between the third and sixth gestational weeks [1]. Any disruption during this period could lead to tracheoesophageal defects.

Tracheal atresia is a very rare entity present in about 1 in 50,000 births [1]. Since its first description in 1900 until 2011, less than one hundred cases were reported [2]. It reflects a partial or complete tracheal absence, with variable extension and location, grouped according to Faro and Floyd classification systems.

In 1962, Floyd et al. defined three types of tracheal atresia: I (proximal trachea is absent and distal trachea presents a communication with the esophagus); II (complete tracheal atresia with normal main bronchi joining at carina with or without a tracheoesophageal fistula); III (complete atresia with separate origin of main bronchi arising from the esophagus) [1]. More descriptive, Faro et al. divided tracheal atresia in seven types, A to G, from complete pulmonary atresia to tracheal stenosis, respectively (Figure 1) [3].

In about 90% of cases, this entity is associated with other defects, which can be part of syndromes as VACTERL (vertebral, anal, cardiac, tracheoesophageal, renal and limb defects) [1].

Prenatal diagnosis is only possible when no tracheoesophageal fistula is present. Without a communication between lungs and esophagus, fluid accumulates in lungs and trachea until the atresic segment, with hyperechogenic lungs and a dilated trachea [1]. When a fistula is present, lung fluid passes through it to the amniotic fluid or the stomach, being imagiology silent.

Neonates with tracheal atresia present with severe respiratory distress soon after birth, and intubation becomes a heavy challenge. Without a tracheoesophageal fistula, death occurs almost immediately. Only an emergent tracheotomy could be life-saving [2].

When a tracheoesophageal fistula is present, after unsuccessful tracheal intubation attempts, esophageal intubation could be the last chance to assure ventilation.

Unfortunately, prognosis of tracheal atresia is poor and this entity is usually lethal. A 2012 review including 49 case reports on this disease, reported an 85% death rate within the first 2 days of life [1].

Figure 1: Classification of tracheal atresia or agenesis. (1) Faro type A: total tracheal and pulmonary agenesis. (2) Faro type B/Floyd type III: complete tracheal agenesis with bronchial origin from esophagus. (3) Faro type C: total agenesis with main bronchi fusing at the carina; presence of a tracheoesophageal fistula. Floyd type II: total tracheal agenesis with main bronchi fusing at the carina; a tracheoesophageal fistula can be present (no image). (4) Faro type D: atresic tracheal segment joining larynx to a normal distal trachea with the presence of a tracheoesophageal fistula. (5) Faro type E/ Floyd type I: proximal trachea atresia with a normal distal segment and a tracheoesophageal fistula. Faro type F: agenesis of the proximal trachea with a normal distal segment without tracheoesophageal fistula (no image). (6) Faro type G: short segment of tracheal agenesis [1].

Case Report

Twenty-five years-old mother, monitored pregnancy, normal fetal ultrasound scans, hydramnios at 34 weeks. Medical history of hypothyroidism and use of tuberculostatic drugs during first trimester.
Male term newborn, normal vaginal delivery in acute fetal distress. Birth weight 2555 g, Apgar score 4/6/7 (1/5/10). Right after birth, abundant oropharyngeal secretions and ineffective respiratory movements, requiring intubation with endotracheal tube at minute 10 with difficult tracheal and nasogastric tubes progression. After intubation, ventilation and saturation have improved. A thoracoabdominal X-ray showed air in stomach and small bowel, with a high tracheal tube extremity, close to nasogastric probe. Diagnosis of esophageal atresia, imperforate anus and adducted thumbs was established.

In the following hours, he presented with progressive difficult and ineffective ventilation. Nasopharyngoscopy showed a collapsed glottic cleft, impeding pediatric fibroscope progression. A contrasted neck and thorax CT scan demonstrated a tracheal atresia of 12 mm, cranial to a proximal tracheoesophageal fistula. After the fistula, trachea presented normal caliber and divided in two main bronchi. It also showed an esophageal atresia with a proximal (C7) and distal tracheoesophageal fistula (to the left main bronchus). Proximal esophagus ended in cul-du-sac, where was found the tracheal tube (T1-T2). Distal esophagus also ended in cul-du-sac (T9-T10), without apparent communication to the stomach (Figure 2) [4].

Echocardiography showed left ventricle output and function reduction, with patent ductus arteriosus with bidirectional shunt and pulmonary hypertension. No aortic coartation.

The newborn evolved within hemodynamic instability requiring fluid and inotropic support and frequent desaturation episodes.

After multidisciplinary discussion involving Anesthesiology, Pediatric Surgery, Otolaryngology, Radiology and Neonatology Intensive Care teams, was decided to plan his transfer to a specialized abroad center with pediatric thoracic surgery to attempt surgery [1].

At 22 h of life he was submitted to emergent gastrostomy and colostomy:

Procedure was performed under balanced anesthesia, with fentanyl perfusion and atracurium. Ventilation was maintained through the esophageal tube, with volume-controlled ventilation with PEEP 5 cmH_2O and good peripheral O_2 saturations (96-100%). Hemodynamic stability was assured with noradrenaline and dopamine perfusions. Procedure was uneventful.

On sixth day of life, newborn presented a severe desaturation episode. Tracheal tube was obstructed and exchanged under videolaryngoscopy; however, new tube maintained no subglottic progression. Several positions in the superior esophagus were attempted. The ineffective ventilation evolved to cardio-respiratory arrest, resulting in newborn death.

Discussion

In pediatric anesthesia, difficult airway is more common at a younger age, presenting in about 0.24% of intubations in infants less than one year [5]. Those patients present a specific challenge, because according to the concrete anatomical defect, recommended approach algorithms may not be enough.

Tracheal atresia is one of the rarest tracheal defects [2], presenting as an unexpected emergency in delivery room, with a rare and unanticipated difficult or even impossible airway management to the anesthesiology and neonatology teams.

This case reports a newborn presenting at birth with a tracheal atresia Faro type E/Floyd type I, with atresia of the proximal trachea and a normal caudal segment of the distal trachea with two tracheoesophageal fistulas and an esophageal atresia (Figure 2).

This Floyd atresia type accounts for the rarest type of tracheal atresia, with an incidence of 15-20% of all cases [2].

As a tracheoesophageal fistula was present, prenatal diagnosis becomes particularly difficult. This case, as the majority of them, was not diagnosed in the prenatal period, despite hydramnios was present in the late gestation weeks, which could alert for tracheoesophageal disorders, present in 70% of cases [1].

Despite esophageal intubation allows temporary ventilation, this would be sooner or later insufficient, and in this specific case, ventilation failure lead to death at the sixth day of life.

Nowadays, surgical and medical management of this condition still offers limited success.

If diagnosed in the prenatal period, further imaging studies can be done during pregnancy to describe in detail the airway malformations, and delivery should be arranged in a specialized center to allow immediate surgical approach after or even during delivery [6].

The only available treatment for tracheal atresia is a complex surgical reconstruction. Depending on the defect the surgical approach can include tracheostomy, esophageal binding or reconstruction, esophagectomy and/or gastrostomy. However most of those solutions present disappointing results with a high mortality rate [6].

Tissue engineering for the reconstruction of the atresic segment could lead to successful therapies in the future [2]. Extracorporeal membrane oxygenation (ECMO) may be a temporary solution until surgery [1].

Initial resuscitation team, namely anesthesiologist and neonatologist, must be fully prepared to manage neonatal difficult airways and recognize those rare syndromes, improving neonatal care and increasing survival rates, as well improving surgical experience and new surgical approaches [2].

In the absence of a visible trachea, esophageal intubation is recommended, expecting that a tracheoesophageal fistula could be present to allow a passage to oxygenate and ventilate the newborn [6].

References