

Diagnosis and Airway Management in a Neonate with Laryngeal Atresia, Tracheal Agenesis and a Broncho-Esophageal Fistula: A Case Report

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Abstract

Tracheal agenesis is a rare congenital anomaly that is usually fatal and occurs in approximately 1:50,000 births. The diagnosis is usually made when there is failure to perform endotracheal intubation in a neonate with severe respiratory distress and absence of audible cry. We present a case of a newborn with suspected trachea-esophageal fistula who presented to the operating room for ligation of the fistula. The diagnosis of laryngeal atresia with tracheal agenesis was made intraoperatively when attempts at surgical ligation of the fistula resulted in difficulty with ventilation and a loss of the capnograph. Ventilation of the lungs was possible with an esophageally placed tube via an esophago-bronchial fistula. This case highlights the importance and need for good communication and teamwork between surgeons, anesthesiologist, radiologist and neonatologists.

Keywords: Laryngeal atresia; Tracheal agenesis; Lung ventilation

Introduction

Tracheal agenesis is a rare congenital anomaly that is usually fatal and occurs in approximately 1:50,000 births with a male: female ratio of 2:1 [1,2]. Several types have been identified by Floyd [3]. Failure to perform endotracheal intubation in a neonate with severe respiratory distress and absence of audible cry are two important clues to the diagnosis of this condition [4,5]. The anesthesia and surgical team may face this challenge either at the delivery of the baby or in the operating room [6,7]. We present a case of a newborn with laryngeal atresia, tracheal agenesis, and a broncho-esophageal fistula. Consent for reporting this case with a view to publication was obtained from the patient's mother.

Case Report

A 33 and 3/7 week gestational age male infant, weighing 1835 grams, was born by spontaneous vaginal delivery to a 19 year old primiparous woman. The neonate was antenatally diagnosed via ultrasound with multiple congenital anomalies, including absent corpus callosum, D-malposition of great vessels, pulmonary stenosis, double outlet right ventricle, ventriculomegaly, midface hypoplasia, spinal anomalies, absent stomach bubble, and 46XY karyotype.

The baby was born with AGPAR scores of 2 at 1 minute, 2 at 5 minutes, 5 at 10 minutes, and 5 at 15 minutes. No respiratory effort or audible cry was detected. Immediately following delivery of the baby, bag-valve-mask ventilation was administered, and the baby was intubated by the neonatology team. Initial intubation was achieved with some difficulty and a 3.0 uncuffed endotracheal tube (ETT) was established with direct laryngoscopy. Tracheal placement was confirmed with an improvement of oxygen saturation, with chest auscultation, and with visual chest rise upon positive pressure via bag-valve-mask ventilation. The infant was ventilated without difficulty and transferred to the Neonatal Intensive Care Unit (NICU) where ETT placement was re-confirmed with capnography and chest auscultation. Shortly after arrival to the NICU, the 3.0 uncuffed ETT was replaced by a 3.5 uncuffed ETT because of an audible leak at peak airway pressures of 20 cm H₂O. Again, tracheal placement after the second intubation was confirmed with capnography and chest auscultation. The fraction of inspired oxy-

gen was adjusted to 0.28 - 1.0 to maintain oxygen saturation 85 - 95% based on pulse oximetry.

An echocardiogram performed on the first day of life revealed a large atrial septal defect, unobstructed total anomalous pulmonary return to coronary sinus, doubles superior vena cava, tetralogy of Fallot, pulmonary artery hypoplasia, and interrupted inferior vena cava with azygous continuation. A chest x-ray on the second day of life revealed an air-filled distended esophagus which was consistent with duodenal atresia. An orogastric tube could not be passed beyond seven centimeters from the gum line. On the fourth day of life, a flexible esophagogastroduodenoscopy was performed at the bedside in the NICU using a 2 mm fiberoptic endoscope by the pediatric surgery team. Additionally, the baby had choanal atresia hence the inability to pass a nasogastric tube. A suspicion of TE fistula was raised based on a bedside flexible endoscopy finding in the NICU which revealed a possible fistula 1 cm above the carina, short esophagus, intra-thoracic stomach and duodenal atresia. Visualization was technically difficult, but it appeared that the patient had a low H-type tracheoesophageal fistula. Since the baby was deemed to be stable from cardiology standpoint, the opinion of the cardiac surgery team was to consider repair of the tetralogy of Fallot and the total anomalous pulmonary venous return when the baby would be 4-5 kg in weight (i.e. feed and grow the baby, pulse oximetry oxygen saturation readings of 75% to 85% were acceptable). Hence, the initial treatment plan was to manage the gastro-intestinal issues.

The baby was transferred to the operating room with the endotra-

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cheal tube in situ on the seventh day of life for ligation of the tracheo-esophageal fistula. Standard monitoring as recommended by the American Society of Anesthesiology was established, and general anesthesia was induced and maintained with an oxygen, air, and sevoflurane mixture and rocuronium was used to facilitate muscle paralysis. Chest auscultation and capnography were used to confirm correct placement of the endotracheal tube. A direct laryngoscopy to confirm endotracheal tube placement at this point was not performed in view of the fact that the endotracheal tube was felt to be optimally positioned and in doing so there was fear of dislodgement of the endotracheal tube. Following this, a right thoracotomy was performed, and the tracheo-esophageal fistula was identified (Figure 1). It was noted that temporary occlusion of the fistula resulted in the loss of ventilation and capnography. Re-evaluation of the anatomy and direct laryngoscopy in the left lateral decubitus position revealed esophageal placement of the endotracheal tube. Therefore, the incision was temporarily closed, and the baby was placed in a supine position in order to facilitate direct laryngoscopy and to correctly place the endotracheal tube.

Direct laryngoscopy with a Miller one blade revealed what appeared to be an epiglottic flap overlying a structure that appeared to be the aryepiglottic folds. At this point, the existing esophageally placed endotracheal tube was removed, and the baby was successfully ventilated via facemask with 100% oxygen. An attempt was made to intubate the trachea correctly. However, the endotracheal tube could not be negotiated past what appeared to be the aryepiglottic folds. Repeated sequential attempts under direct laryngoscopy included 3.0, 2.5, and 2.0 uncuffed endotracheal tubes. All attempts at placement with direct laryngoscopy failed with the inability to negotiate the endotracheal tube past the glottis. Mask ventilation was easily continued during these

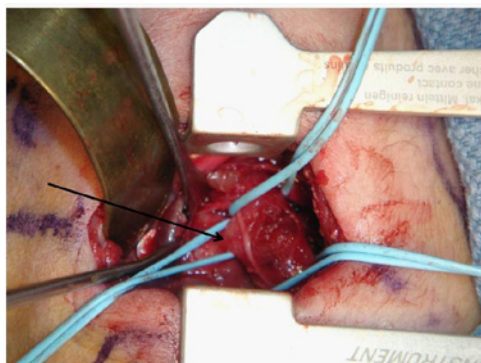


Figure 1: Intra-operative identification of the suspected TE fistula.

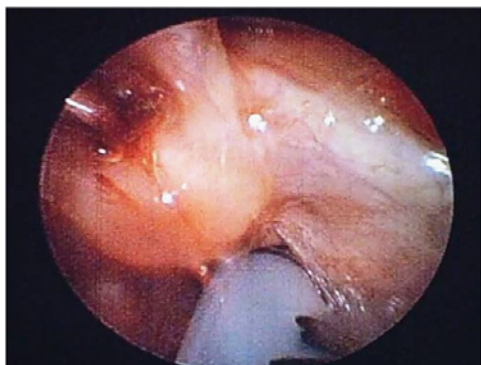


Figure 2: Flexible fiberoptic bronchoscopy view of the upper airway showing the endotracheal in the esophagus and an atretic larynx anteriorly.

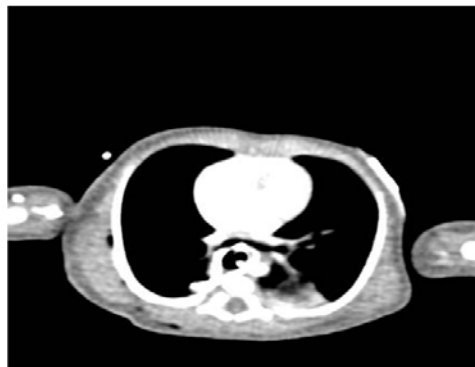


Figure 3: CT scan of the lungs the level of the carnia, showing the fistulous connection from esophagus to short carnia.

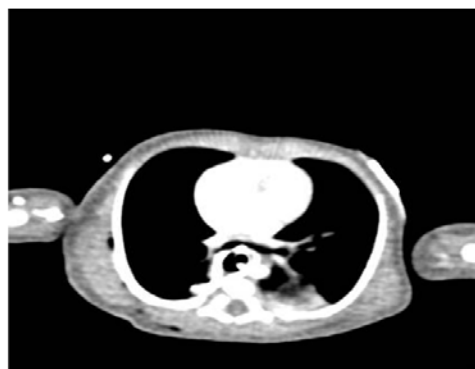


Figure 4: Autopsy specimen of the lungs with the esophagus and stomach the band identifies the broncho-esophageal / carinoesophageal fistula, there is complete absence of the trachea.

attempts and the baby's oxygen saturations were maintained 80-94% as measured by pulse oximetry.

The surgical team subsequently performed flexible and rigid bronchoscopy which confirmed the suspicion of laryngeal atresia (Figure 2). At this point the esophageal intubation and ventilation of the lungs via the fistula was reestablished. The patient was then placed in the left lateral decubitus position to facilitate closure of the incision. Following discussion with the radiologist, the baby was transported for computed tomography scanning to evaluate the presence of a trachea for a possible tracheostomy. Adequate ventilation of the lungs continued via the esophageally placed endotracheal tube throughout the process.

The computed tomography (CT) scan revealed a very short fistulous connection from the distal esophagus to a small carina which immediately separated into the left and right main stem bronchi (Figure 3). There was an absence of an identifiable trachea. Numerous other complex congenital cardiac anomalies were seen.

The baby was transported back to the Neonatal Intensive Care Unit, and the prognosis was discussed with the mother. The decision was made to electively extubate the baby. The baby deceased within 24 hours. A complete autopsy revealed agenesis of the trachea and a larynx that measured 0.7 cm in length. The presence of a bronchial-esophageal fistula was confirmed (Figure 4).

Discussion

We have reported a case of a newborn with suspected tracheo-esophageal fistula and duodenal atresia that was intraoperatively found

to have tracheal agenesis with laryngeal atresia. Ventilation of the lungs was possible via an esophago-bronchial fistula. The suspicion of an esophago-bronchial ventilation pathway was raised when attempts at surgical ligation of the fistula resulted in difficulty with ventilation of the lungs.

Since the patient maintained respiratory stability that was commensurate with the comorbidities or coexisting diseases for several days after intubation, confirmation of the endotracheal tube position by direct laryngoscopy was not made in the operating room. Auscultation, capnography, and oxygen saturation by pulse oximetry were used for confirmation of the endotracheal tube upon arrival of the baby in the operating room. Perhaps if direct laryngoscopy for confirmation of laryngotracheal placement had been performed prior to the commencement of the surgical procedure, thoracotomy may have been deferred, although it may not have changed the course of the disease and the outcome. However, in our case, direct laryngoscopy was not performed in view of the precarious positioning of the ETT in relation to the suspected fistula, for fear of dislodgment of what was thought to be an optimally placed ETT. Difficulty ventilating the patient in the operating room upon occlusion of the presumed tracheoesophageal fistula raised the possibility of an esophageal intubation. The ultimate outcome was not affected by this delay in diagnosis, though prompt diagnosis may have prevented the patient from undergoing a thoracotomy. Fiberoptic endoscopy confirmed the presence of laryngeal atresia and tracheal agenesis in the operating room.

Available literature suggests various types of tracheal agenesis [7,8]. Based on Floyd's classification, our patient appears to fit into Floyd Class 2 i.e. atresia of the complete trachea with a normal carina and main bronchi and with or without carino-esophageal fistula [1]. There has been a report of a newborn with laryngeal and tracheal agenesis where the baby was intubated and later found to have laryngo-tracheal atresia [9]. Suspicion of a laryngeal atresia and tracheal agenesis (causes of Congenital High Airway Obstruction Syndrome, CHAOS) can be made by antenatally based on ultrasound findings of fetal ascites, fluid in the upper airway or enlarged and hyperechogenic lungs [10]. However these signs may be absent in the presence of a tracheo-esophageal or a broncho-esophageal fistula (as in our case) because the fluid from the lungs can pass into the stomach via the fistula. Hence, in our case, an antenatal ultrasound failed to raise concerns for a possible laryngeal atresia and tracheal agenesis. Suspicion for a possible trachea-esophageal fistula was based on clinical presentation and endoscopic findings after birth. Antenatal suspicion of tracheal agenesis may be confirmed by fetal magnetic resonance imaging (MRI) scanning [10]. Patients with CHAOS may be suitable for an EXIT (Ex-utero intrapartum therapy) procedure. Use of the EXIT strategy has been described in a patient with complete tracheal agenesis where the mediastinum was explored and the baby intubated at the level of the right main stem bronchus [11]. An isolated laryngeal atresia, if diagnosed at birth, would warrant an emergent tracheostomy. An urgent computed tomography scan in our patient was justified as it had implications on further management of the patient in terms of the ability to perform a tracheostomy.

Our patient's tracheal agenesis was not antenatally diagnosed. While tracheo-esophageal fistula was suspected, several issues related to the clinical history suggested tracheal agenesis prior to the surgical procedure. In retrospect, the Neonatal Intensive Care Unit team described two important clinical clues to this condition. The orogastric tube that was passed could not be placed on active suction without losing the ability to ventilate the patient. Second, endotracheal suctioning was significant for copious amounts of gastric secretions. This was attributed

to passage of gastric contents through a suspected trachea-esophageal fistula rather than esophageal intubation per se. Although in our case oxygen saturation as measured by pulse oximeter varied from 80% to 95%, the number was deemed to be acceptable because the patient also had a mixed congenital cardiac lesion. In reported literature, attempt at surgical reconstruction may prolong survival in these babies [12,13]. Esophageal intubation to maintain airway with gastrostomy and distal esophageal banding has also been reported as one of the options to prolong survival in this in a baby with Floyd type 2 tracheal agenesis [13]. Fuchimoto et al. have reported a case of a baby with Floyd type 1 tracheal agenesis (agenesis of the proximal trachea with a normal caudal segment of the distal trachea and a trachea-esophageal fistula) who was successfully managed with esophageal banding to substitute esophagus for trachea and a cervical esophagostomy with a gastrostomy tube for feeding [14]. Perhaps, application of regenerative medicine and tissue engineering with amniotic fluid cells for laryngotracheal reconstruction in case of antenatally diagnosed laryngotracheal agenesis may be lifesaving, although this is still in its early stages of research and clinical application [15].

In summary, we have presented a case of a neonate who was intraoperatively diagnosed tracheal agenesis and laryngeal atresia. These cases pose a diagnostic, clinical management and ethical challenge. A high index of suspicion for tracheal agenesis and laryngeal atresia must be raised when a baby presents with a tracheo-esophageal fistula and multiple congenital anomalies. Although the clinical presentation may be similar, the treatment and outcome is different.

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