Oesophageal Intubation and Ventilation as Initial Airway Support of Newborn Infant With Tracheal Agenesis

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SUMMARY

Tracheal agenesis is a rare congenital airway anomaly which presents as an airway emergency at birth. We report a case of late premature Chinese infant with tracheal agenesis type II (by Floyd's classification) who presented with severe respiratory distress at birth. He had multiple failed attempts at intubations with accidental oesophageal intubation and ventilation. Tracheal agenesis with tracheo-oesophageal fistula was suspected from an emergency optical laryngoesophagoscopy done. The infant was subsequently stabilized on oesophageal ventilation. The diagnosis was confirmed on CT scan and parents were counseled regarding the poor outcome and decided for withdrawal at day 7 of life.

KEY WORDS:
Tracheal agenesis; Tracheal atresia; Newborn

INTRODUCTION

Tracheal agenesis is a rare congenital airway anomaly with an incidence of about 1 in 50000 births[1]. Infant with tracheal agenesis usually presents with severe respiratory distress at birth with difficult airway management. Therefore a high index of suspicion is necessary when an infant presents with respiratory distress at birth with no audible cry and endotracheal intubation is unachievable. Oesophageal intubation may allow ventilation of the lungs while trying to establish the diagnosis. The overall prognosis remains poor and no effective surgical intervention is currently available in Malaysia.

CASE REPORT

A late preterm infant at 35 weeks gestation with a birth weight of 2356 g was born via spontaneous vaginal delivery. There was an antenatal history of maternal gestational diabetes mellitus on diet control complicated with polyhydramnios. However, no detailed antenatal ultrasound was done.

Following an uncomplicated delivery, the baby was unexpectedly depressed at birth with poor Apgar score of 1 at 1 minute and 5 at 5 minutes of life. He was started immediately on bag and mask ventilation. Repeated attempts at oral endotracheal intubation failed even with the smallest size endotracheal tube of 2.5 mm. In the process, the oesophagus was accidentally intubated which improved and maintained the respiratory status. Emergency direct laryngoesophagoscopy examination by the otorhinolaryngologist revealed a hypoplastic laryngeal inlet with a small glottis opening. A stylet passed through the glottis opening could not go beyond 2 cm. There was an opening at the oesophagus about 3 mm into the trachea just above the carina. The infant was suspected to have tracheal agenesis with tracheo-oesophageal fistula.

Initial blood gas showed severe mixed respiratory and metabolic acidosis. This was gradually corrected after oesophageal ventilation. Examination revealed a single umbilical artery but otherwise no other congenital abnormality. Bedside echocardiography and ultrasound kidneys were normal. Once the infant was stable, a repeat laryngoesophagoscopy was done the next day which showed a hypoplastic laryngeal inlet with a cleft larynx ending in a blind pouch a few millimeters below the vocal cords. Both the endotracheal tube and nasogastric tube were within the esophagus (Figure 1). There was a fistula opening at the antero-lateral oesophageal wall 12 centimeters from the incisors which led to the carina and left and right bronchus.

CT thorax was done at day 5 of life to delineate the airway anatomy and to help decide whether any surgical intervention was possible. The contrast-enhanced CT thorax confirmed subglottic tracheal atresia with absence of lower cervical and thoracic trachea. A tracheo-esophageal fistula was identified with both bronchi originating from a flattened, horizontally orientated carina which was in direct communication with the oesophagus (Fig 2). Based on the CT thorax, isolated Type II tracheal agenesis was diagnosed.
The condition was deemed inoperable following discussion with the otorhinolaryngologist and the paediatric surgeon. The parents were counselled regarding poor prognosis and a decision for withdrawal of life support was done at day 7 of life.

DISCUSSION

Prenatal diagnosis of tracheal agenesis (TA) is rare and only possible in the absence of tracheo-oesophageal fistula (TOF) as the patients would present with classical congenital high airway obstruction syndrome (CHAOS). CHAOS is characterized by enlarged hyperechogenic lungs, fluid filled dilated trachea and bronchi, with an absent flow in the trachea during fetal breathing[2]. These signs are absent in the presence of TOF as lung fluid can be drained through the TOF, allowing the lungs to appear normal ultrasonographically. However, polyhydramnios and other

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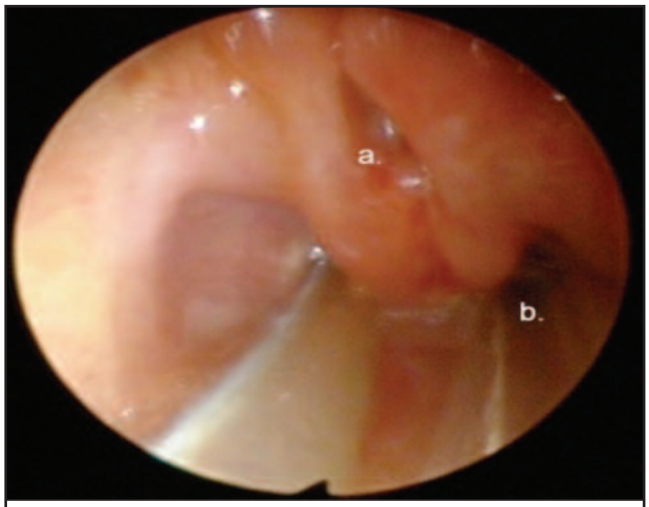
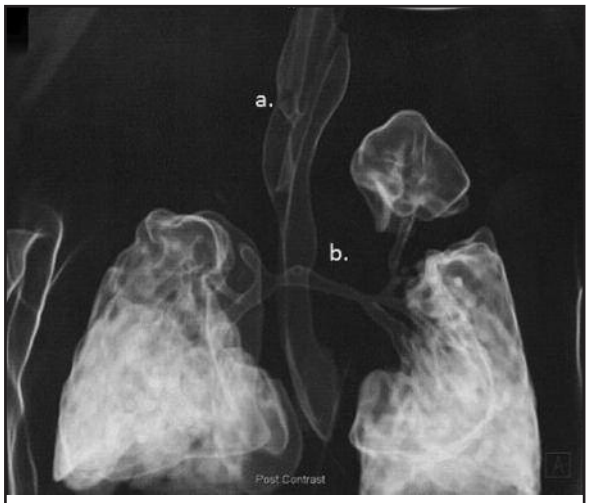


a. Hypoplastic laryngeal inlet with cleft larynx
b. Oesophageal opening with both the endotracheal tube and nasogastric tube entering through

Fig. 1: Direct laryngoesophagoscopy of the larynx.



a. Subglottic tracheal atresia
b. Tracheo-oesophageal fistula with both bronchi originating from a flattened carina

Fig. 2: 3D reconstruction of the upper airway.

congenital malformation could alert the clinician regarding potential tracheal problems [3].

A high index of suspicion is necessary for the prenatally undiagnosed infant who presents at birth with respiratory distress. Infants with TA are characteristically premature, have low birth weight (<2500g) and poor Apgar score below 7 at 5 minutes. They usually present with respiratory distress at birth with breathing movement but poor air entry, no audible cry and failed endotracheal intubation [3].

Oesophageal intubation and ventilation (accidental or intended) may provide temporary improvement of respiratory status in infants with TA and TOF. Diagnosis can be ascertained by laryngoesophagoscopy but CT scan is the modality of choice to delineate the anatomy of the upper airway. Tracheal agenesis is classified into 3 anatomical subtypes according to Floyd classification [4]. Type I is characterised by agenesis of the proximal trachea and presence of a distal tracheo-oesophageal fistula. In type II, there is a complete absence of distal trachea but with a presence of normal bifurcating bronchi. While in type III, the main bronchi arise independently from the oesophagus. Isolated type II tracheal agenesis as in this case is rare and up to 94% are associated with other congenital abnormalities such as congenital cardiac, genitourinary, gastro-intestinal, pulmonary, central nervous system and musculoskeletal anomalies.

Oesophageal ventilation is only possible in the presence of tracheo-oesophageal fistula or broncho-oesophageal fistula. In type I, the endotracheal tube can be threaded through the distal tracheal stump using flexible fibrescopy. In type II and III, oesophageal intubation with a relatively large endotracheal tube can be placed at the level just above the bronchial communication to maintain the airway albeit with more difficulty. A nasogastric tube should also be inserted for continuous gastric drainage. Use of oesophagus as tracheal substitute poses multiple problems. The collapsible nature of the oesophagus increases the risk of hypoxic events, accumulation of secretions increases risk of chest infections, and erosion of the wall can result in bleeding and air leak [5]. Oesophageal ventilation is only a temporary measure and surgery is mainly palliative. Tracheal reconstruction with prosthetic material and tissue engineering has not yielded satisfactory results.

CONCLUSION

The overall prognosis of TA remains poor and most die in the newborn period with no effective surgical treatment yet available. Oesophageal intubation and ventilation may provide temporary improvement in the respiratory status so as to provide some extra time for confirmatory diagnostic procedures, family counselling and agreed management plan in view of the dismal prognosis of this condition.

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