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Case Report

Unexpected Failure Intubation in a Preterm Newborn with Tracheoesophageal Fistula and Duodenal Atresia: A Case Report of Congenital High Airway Obstruction Syndrome Complicated with Esophageal Atresia

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Abstract Congenital High Airway Obstruction Syndrome (CHAOS) is a rare and critical neonatal condition characterized by upper airway defects, often accompanied by tracheoesophageal fistula (TOF). Concurrent esophageal atresia (EA) is exceedingly rare, complicating intubation efforts. We present a case of a preterm male neonate with CHAOS, TOF, EA, and duodenal atresia, who experienced unexpected failure in both tracheal and esophageal intubation despite prior TOF confirmation. The case underscores the importance of thorough antenatal examination for early diagnosis and highlights the need for innovative tracheal replacement strategies. Timely and appropriate airway management is crucial for improving prognosis in such cases. This report aims to raise awareness and guide clinical preparedness for managing infants with CHAOS and EA. Future research should focus on identifying superior tracheal replacement options.

Key words *Congenital High Airway Obstruction Syndrome; Tracheal atresia; Tracheoesophageal fistula*

Introduction

Congenital High Airway Obstruction Syndrome (CHAOS) is a rare and extremely fatal congenital abnormality and emergency for neonate. It consists of several defects in upper airway. It may accompany with or without tracheoesophageal fistula (TOF) when having antenatal examination. Without timely and proper ventilation, the prognosis will not be ideal. The diagnosis maybe often confirmed by autopsy. We report a case of a male preterm newborn with CHAOS. Unexpected failure of trachea and esophageal intubation for resuscitation and ventilation temporarily happened, although TOF had been confirmed. The diagnostic methods, potential treatment options and prognosis are discussed. This case report is valuable in increasing awareness of this rare condition and will help us in being better prepared in managing these children. Future studies should aim at finding the optimal replacement for the tracheal.

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Case Presentation

A 28-year-old gravida 1 para 1 was referred to our hospital for common antenatal examination. The fetus was found to have duodenal obstruction, persistent left superior vena cava, and polyhydramnios by ultrasound at the gestation age (GA) of 24-weeks. The fetus also suspected with TOF and bilateral bronchial atresia (Figure 1A). A

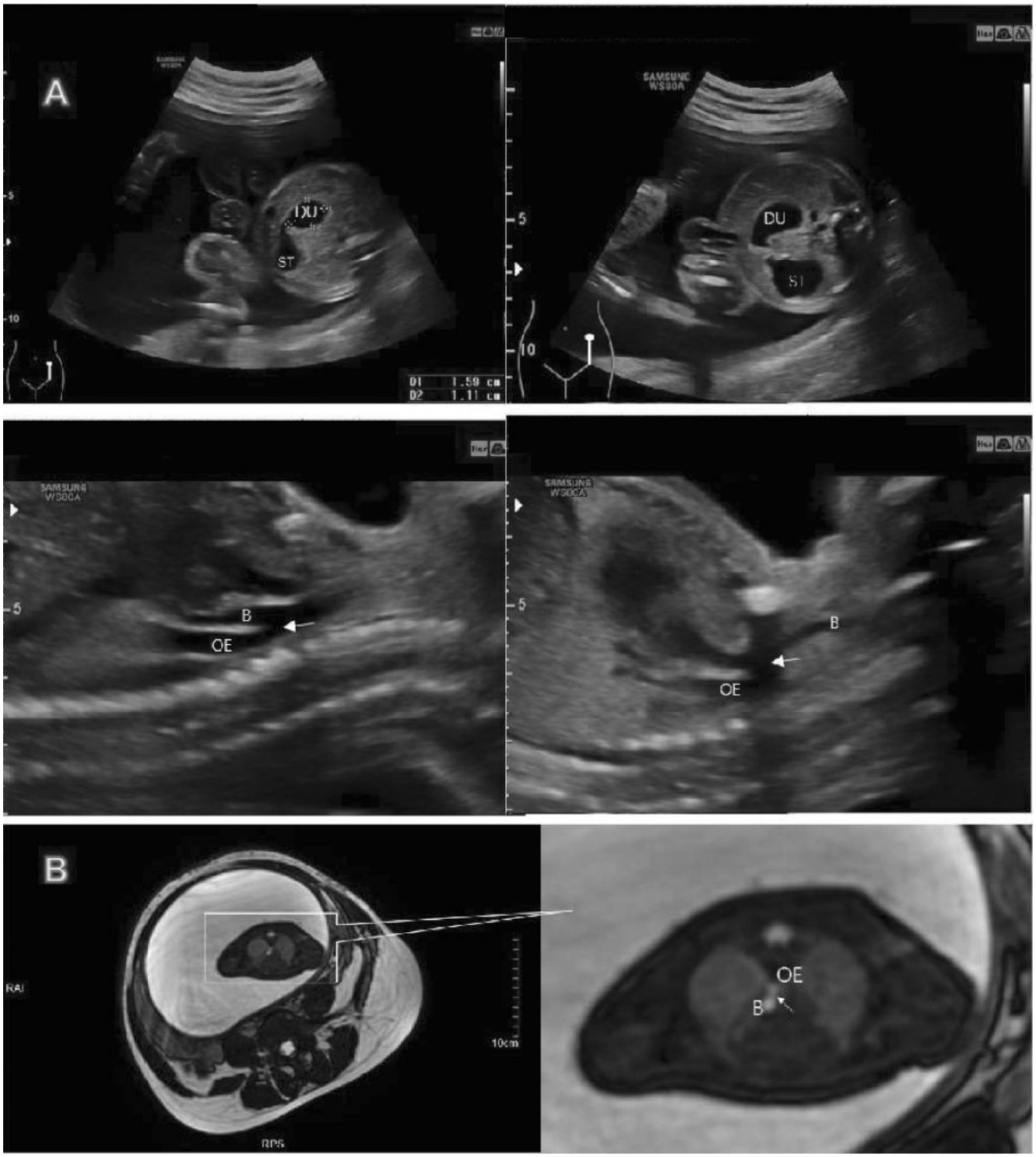

Figure 1 Prenatal ultrasound and magnetic resonance imaging. (A) Ultrasound showed findings of tracheoesophageal fistula and suggestive bilateral bronchial atresia (DU: duodenum; ST: stomach; B: bronchus; OE: esophagus; White arrow: tracheoesophageal fistula). (B) Magnetic resonance imaging showed findings of tracheoesophageal fistula (B: bronchus; OE: esophagus).

fetal magnetic resonance imaging (fMRI) was completed at GA of 29-week, showing proximal duodenal dilatation with severe distal duodenal stenosis, polyhydramnios, and no bilateral bronchial atresia (Figure 1B). Meanwhile, the karyotype and micro array analyse of the fetus amniotic fluid culture showed no obvious abnormality. So this mother decided to go on pregnancy. Due to maternal respiratory distress and a high level of amniotic fluid index, we drew out about 1300 ml and 1500 ml of amniotic fluid by ultrasound-guided amniotic fluid reduction at GA of 31-and 34-week respectively, and gave one course of dexamethasone to promote fetal lung maturation. At GA of 35-week, a Caesarean section was given because of cardiac insufficiency (compression type), polyhydramnios and multiple fetal malformation. A male preterm neonate weighed 2760 g (weight for gestational age, p50-75) delivered with approximate 8300 ml of amniotic fluid. Apgar score was 7 at 1 minute and 8 at 5 minutes. Since there was no indication of severe airway malformations before birth, the baby was only given routine resuscitation after birth. The baby was transferred to our neonatal intensive care unit with severe dyspnea and cyanosis, at 15 minutes after birth. Physical examination showed absence of spontaneous breathing movements, bradycardia and extra digits of right hand. Mask ventilation and endotracheal intubation started immediately. We exposed glottis, and tried to intubate using endotracheal tubes with diameter from 3.5 mm to 2.0 mm, as well as a stylet which is modeling the tube for easier intubation, thinner than endotracheal tube. However, no matter what size of endotracheal tubes or stylet was used, failed to pass through the glottis. During the attempt of inserting endotracheal tubes, mask ventilation with high pressure settings was attempted to maintain the respiratory status of the baby. Under this situation, we considered that this baby should have the possibility of CHAOS with severe tracheal stenosis or atresia. Previous antenatal examination suggested a TOF exist, so we attempted to intubate through the esophagus. Nevertheless, the intubation was unsuccessful with any size of tubes and the stylet. We wondered whether the baby could maintain ventilation through the trachea or the esophagus using mask ventilation as intubation could not be performed through either the trachea or esophagus. Chest X-ray indicated that the tracheal stylet tip was located at the glottic opening level (Figure 2A). Neck and chest computed tomography scan and airway reconstruction showed that there was a suspicious blind end at the distal of the main trachea, while the esophagus above TOF was not shown (Figure 2B). Unfortunately, after a period of prolonged hypoxia, the

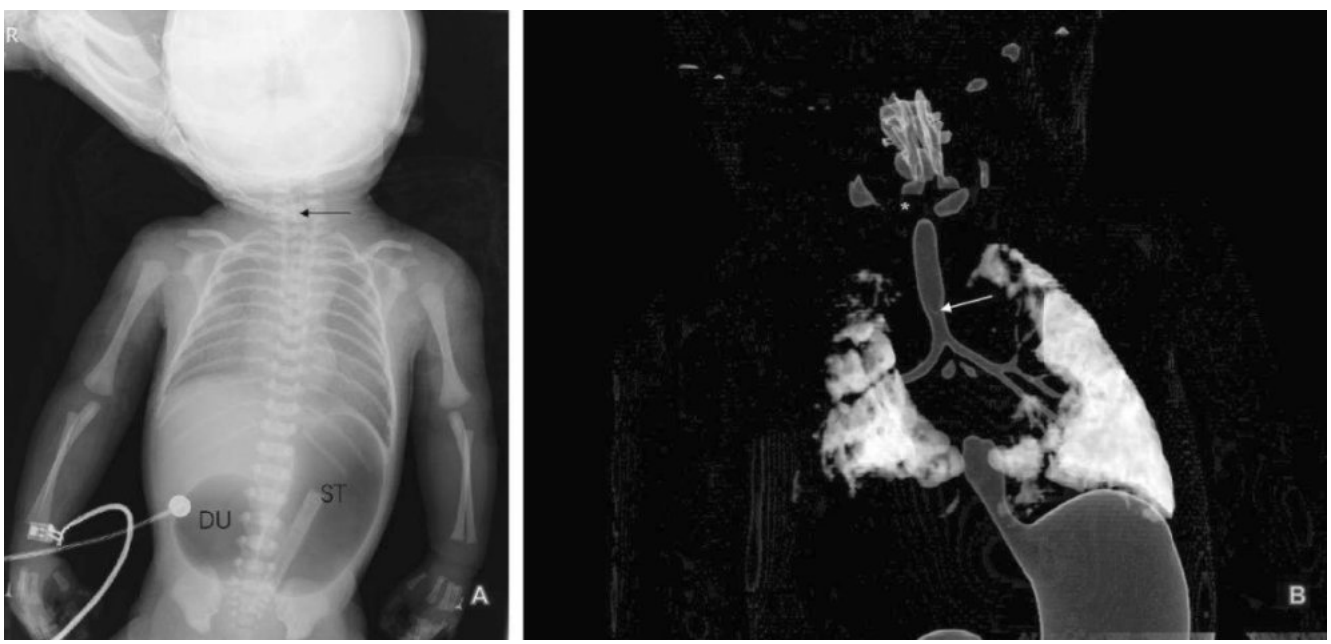

Figure 2 The X-ray and computed tomography scan of the baby showing the tracheal atresia. (A) The X-ray showing the tracheal atresia. and duodenal atresia (Black arrow: the tracheal stylet was located at the glottic opening level; DU: duodenum; ST: stomach) (B) Neck and chest computed tomography scan showing the tracheal atresia and tracheoesophageal fistula (White arrow: there was a suspicious blind end at the distal of the main trachea, while the esophagus above the trachea esophageal fistula was not shown).

blood gas measurement at 30 minutes after birth showed pH 6.529, PO_2 31.8 mmHg, PCO_2 > 150 mmHg, and lactate 17 mmol/L. The parents were counselled regarding poor prognosis and a decision of withdrawal of life support was made at 1 hour of life. Nearly 2 hours after birth, this baby died of respiratory failure. The autopsy showed severe tracheal stenosis, esophageal atresia, TOF, and duodenal atresia, which confirmed our previous inference. Postnatal array-based comparative genomic hybridization (array-CGH) showed no microdeletion or microduplication in the genome.

Discussion

CHAOS is a rare and extremely life-threatening emergency in delivery room. It consists of several defects in upper airway, such as laryngeal atresia, laryngeal or tracheal webs, laryngeal cysts, tracheal atresia, subglottic stenosis or atresia and laryngeal or tracheal agenesis. The mortality is quite high without timely and proper management of the airway. Even if certain procedures are taken to save lives, there may be hypoxia injury of multiple organ. Without a thorough antenatal examination, resuscitation will be challenging. Diagnosis should be suspected in cases of severe respiratory distress, sometimes with no audible cry, and when there is a mechanical blockage preventing intubation.[1]

One of the most unusual and serious problem of CHAOS is tracheal atresia (TA).[2] Since the first reported case of this malformation described by Payne in 1900,[3] over 150 case reports of TA have been published in literature. Only a few cases of neonates with TA were successfully resuscitated. It is estimated the incidence is 1 per 50,000 newborns.[1] TA can present as a single organ malformation, but in 93% cases, multiple congenital abnormalities are encountered and can be part of the VACTERL (Vertebral Defects, Anal Atresia, Cardiovascular Defects, Tracheoesophageal Fistula and/or Esophageal Atresia, Renal Defects, and Limb Defects) association or TARCD (Tracheal Agenesis/Atresia, Radial Ray Defects, Complex Congenital Cardiac Abnormalities, and Duodenal Atresia) association. A concomitant tracheo- or broncho-esophageal fistula is often seen, and its presence allows for resuscitation and ventilation through an esophageal intubation temporarily. However, despite the advances in fMRI and ultrasound technology in recent years, antenatal accurately diagnosis of TA remains challenging. Some clinicians even found TA after a second reading of MRI images.[4] If a TOF is present, polyhydramnios and other congenital malformations on ultrasound should alert clinicians of potential tracheal problems.[5]

In anticipation of difficult airway, the ex-utero intrapartum treatment (EXIT) procedure is an effective management strategy to ensure a safe delivery.[6] During the EXIT-procedure, either by establishing temporary surgical airway or applying extracorporeal membrane oxygenation (ECMO) cannulation can be performed to bridge to a later surgical repair.[7] Emergency tracheotomy has been performed in some cases of tracheal agenesis, but the procedure is rarely successful and complex.[8]

Therefore, some form of tracheostomy solution has to be found. Its success depends on the length of the distal trachea. In most cases long-term solutions for tracheal agenesis remained very limited.[9] In summary, the prognosis of tracheal agenesis is very poor, even though survival with experimental therapies has been reported in literatures.[10] Patient management varies according to the center and the available facilities; however, the main aim of surgical intervention in emergent patients is to secure the airway and provide sufficient oxygen.

Conclusion

CHAOS is a rare and extreme life-threatening emergency in delivery room. TA is one of the most unusual and serious problem. A thorough antenatal examination may provide important information for resuscitation. The mortality is depending on timely and proper management of the airway. Once the diagnosis is confirmed, EXIT or tracheotomy should be accepted by experienced clinical experts if local medical resources allowed.

Declaration of Interest

No conflict of interest involved.

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