Stumbled across a rare entity tracheal agenesis – A lesson learned

Sir,

Complete tracheal agenesis (TA) is a rare congenital entity and occasionally life-sustaining when allied with a tracheo-esophageal or broncho-esophageal fistula. The male-to-female preponderance is 2:1, with a prevalence of less than 1:50,000.[1] There exists a dearth of cases reported. Moreover, worldwide approximately <200 cases have been documented since 1900 when Payne for the first time described this disorder.[2] The prognosis is grave and most of the neonates succumb immediately after birth. Antenatal diagnosis is arduous and postnatal clinical pointers; severe asphyxia with intense respiratory efforts, silent cry, and failed endotracheal intubation with a non-palpable tracheal ring should arouse TA speculation.

We present a neonate referred to our hospital, who landed up in the emergency unit with severe birth asphyxia, failure to cry, and a heart rate of 50/min. Immediately

cardiopulmonary resuscitation was started. Neonate was intubated with an endotracheal tube 3.0 mm ID. After shifting to our hospital and initial stabilization, an umbilical artery and vein catheter was inserted under aseptic precaution, and a mean arterial pressure of 31 mm Hg was noted. Intravenous fluid was started along with 10% dextrose and inotropes infusion was started with injection Dopamine and Dobutamine (20 μg/kg/min). After 20 min approximately, continuous bubbling was observed from the orogastric tube was observed and a fall in saturation with gastric distension was observed. The baby was reintubated for suspicion of tube block and documented no laryngeal opening seen or single opening for the laryngeal/esophagus and a call was made to the senior anesthesiologist for the difficult intubation. At that time, sub-glottic stenosis was suspected and the ENT team did rigid endoscopy and found a normal larynx leading to the trachea that end in a blinded pouch, and suspicion of TA was suspected. Although we also thought tracheostomy but trachea was not palpated below the cricoid cartilage. Thereby tube was inserted in the esophagus and bilateral chest rise was noted. Saturation started improving and the child was transitionally stable [Figure 1]. A plan of CT scan followed by shifting the patient to the operating room for further evaluation and palliative lifesaving surgery was planned. Finally, after 30 min, the neonate again had arrested and could not be revived.

Prenatal diagnosis is difficult and postnatal diagnosis is usually made during a respiratory emergency. This anomaly is often amalgamated with respiratory, cardiovascular, and gastrointestinal abnormalities. The compatibility with life is exclusively possible only with the presence of tracheo-esophageal or broncho-esophageal fistula in this anomaly.[3]

For instantaneous management of severe distress, we recommend esophageal endotracheal intubation to impart air aisle and apprehend the level of a defect by the cautious use of roentgenography and contrast material. Immediate tracheostomy may be beneficial in Type 1 TA. Long-term management is extremely challenging, various anastomosis and grafts (homologous/synthetic) have been tried, albeit the results are unsatisfactory.[4,5] The long-term survival of TA was deemed an effective surgical intervention. In the future, tissue engineering may lead to the probability of effective surgical repair and better survival. Therefore, well-evolved skills for the difficult neonatal airway are highly warranted, and cognizance of various airway possibilities will enhance the chance of neonatal survival in this anomaly. Prenatal diagnosis by magnetic resonance imaging can aid in making decisions in an unexpected life-threatening emergency in the postpartum period, especially when ventilation is difficult from the beginning. Our opinion is that we need to venture effective resuscitation attempts in the improvement and development of improved surgical techniques, which should in the time ahead resolve or abate these odd congenital anomalies.

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Conflicts of interest
There are no conflicts of interest.

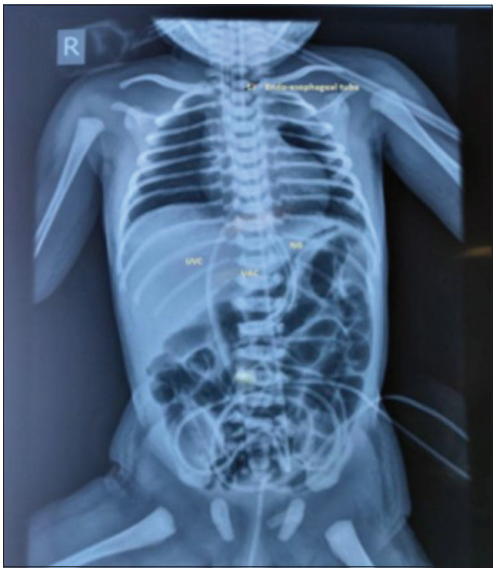

Figure 1: Roentgenography. UVC: umbilical vein catheter; UAC: umbilical artery catheter

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