[NM-236] Unexpected Difficult Intubation in the Operating Room: A Case Report of Tracheal Agenesis

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Tracheal agenesis(TA) is a rare congenital malformation with an incidence of 1:50,000. It is part of the Tracheal Atresia, Cardiac and Renal anomalies, Duodenal atresia(TACRD) association, but is also seen with Vertebral anomalies, Anal atresia, Cardiovascular anomalies, Tracheoesophageal fistula, Esophageal atresia, Renal and Limb anomalies(VACTERL). Floyd et al. classified TA into three types(Fig. 1). Despite polyhydramnios in most pregnancies, prenatal diagnosis is uncommon. Diagnosis is usually made minutes after birth. Typically, the infant presents with cyanosis, severe respiratory distress, poor gas exchange, absence of an audible cry, and difficult or impossible intubation in the delivery room(DR). We present a case where diagnosis was missed in the DR and made in the operating room(OR).

Our patient was a 1-day-old, 1.8 kg monochorionic diamniotic twin delivered at 34 weeks by cesarean section. She was prenatally diagnosed with anhydramnios, bilateral hydronephrosis, cloaca malformation, and double outlet right ventricle(DORV). Her twin was normal. She required CPR in the DR and was intubated on a second attempt. An orogastric tube was placed and chest x-ray showed the endotracheal tube(ETT) in the mid trachea. Transthoracic echo confirmed DORV, large unrestricted VSD, and overriding aorta. Oxygen saturations in the mid 80s were attributed to her cardiac defect. Other anomalies included absence of right thumb, right foot polydactyly, and high imperforate anus. She was taken to the OR for vesicostomy and divided colostomy. On transfer to the OR table, oxygen saturations declined despite ventilation with 100% oxygen. The ETCO2 waveform was inconsistent with tracheal intubation, so the ETT was removed and mask ventilation improved oxygen saturations. Direct laryngoscopy showed Grade 1 view, but despite multiple attempts, even a 2.0 uncuffed ETT would not pass. Rigid bronchoscopy by the otolaryngologist revealed complete TA just distal to the vocal cords. Neck exploration for tracheostomy confirmed absence of trachea down to the manubrium. Esophageal reintubation was performed to maintain oxygen saturations. Extracorporeal membrane oxygenation was proposed, but considered a heroic measure with her cardiac defect. After multispecialty and multi-institutional consultations, there was consensus that the defect was incompatible with life, and care was withdrawn.

In this case, unrecognized esophageal intubation in the DR led to delayed diagnosis of TA. Advancement in prenatal diagnosis would be ideal, but early recognition in the DR is essential for optimal management. Tracheal reconstruction has been described but long-term survival has not yet been achieved.

References

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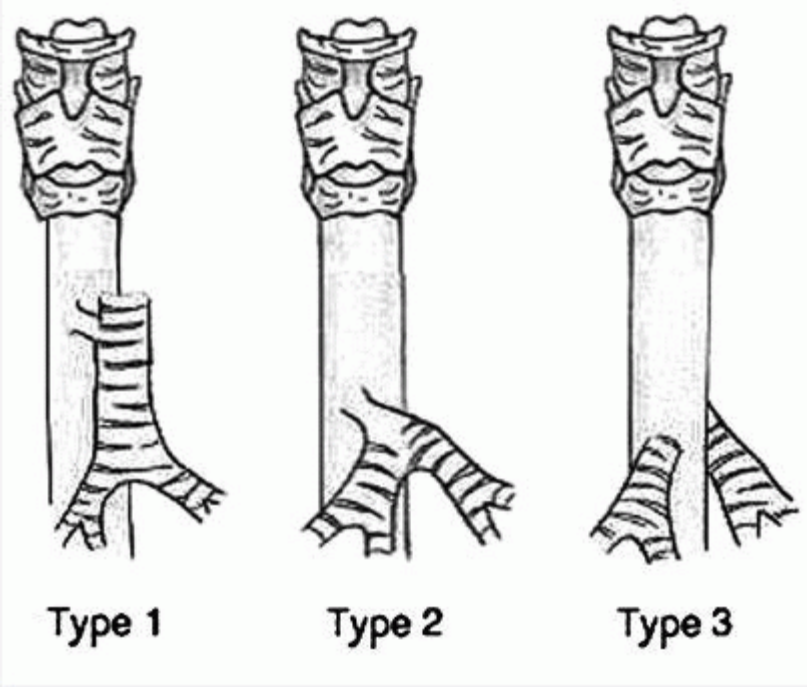
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