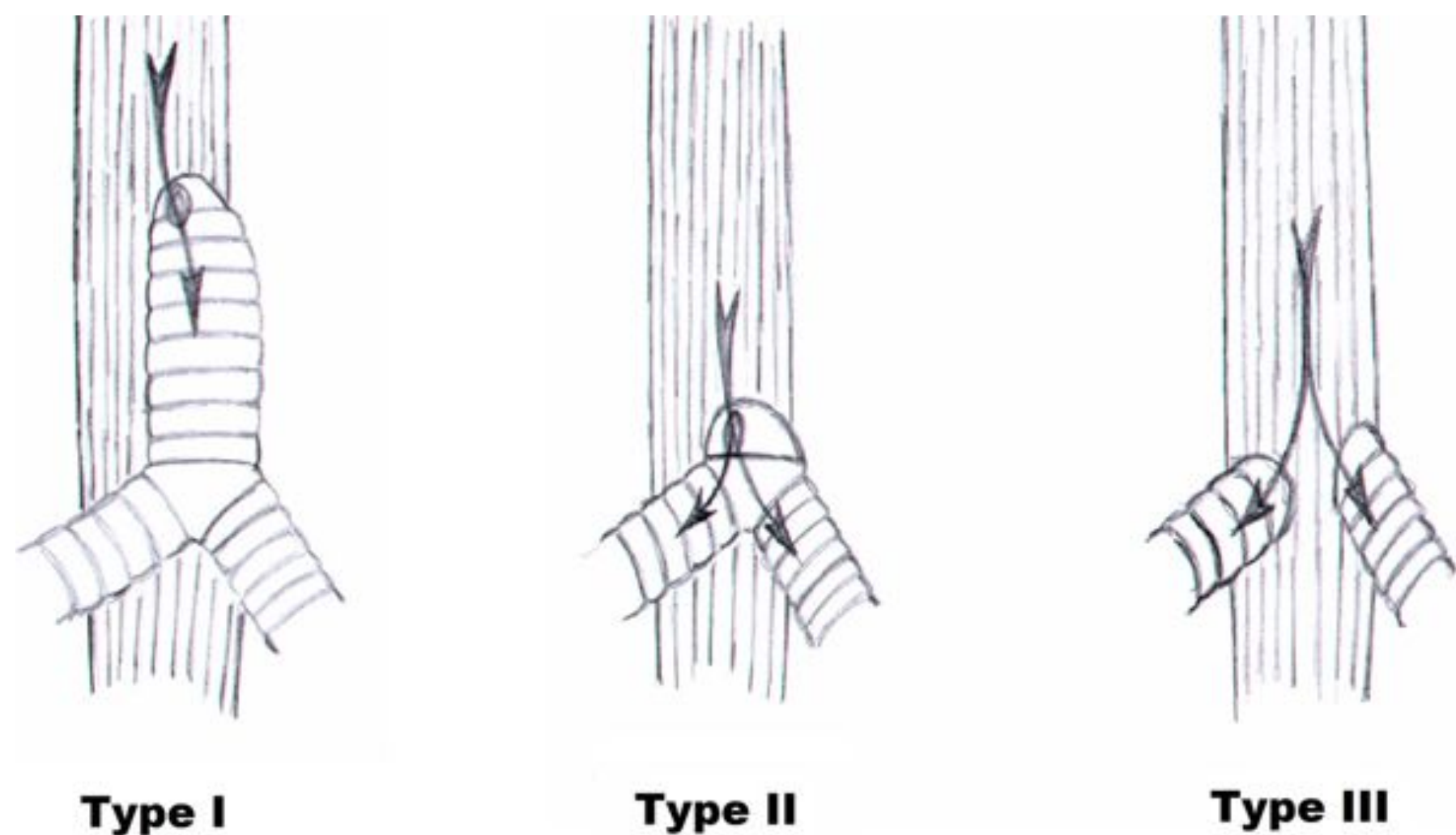


Introduction

Tracheal agenesis is a rare congenital anomaly with an incidence of about 1 in 50,000 births, with nearly half of the cases associated with in-utero polyhydramnios and 64% resulting in premature birth. Tracheal agenesis often occurs as part of the VACTERL association. Floyd's classification distinguishes between three types of tracheal agenesis.



Postnatal diagnosis depends on clinical signs such as respiratory distress and difficult intubation. In most cases, diagnosis is confirmed with a CT scan and laryngo-bronchoscopy. Although tracheal agenesis can be a fatal diagnosis, the treatment includes esophageal reconstruction and the use of tracheal prostheses.

Here, we discuss a case in which intentional esophageal intubation was successfully used to temporize an airway in a neonate with Floyd Type II tracheal agenesis.

Case Description

- Baby girl born at 37 weeks of gestation presented with apnea and hypotonia after vacuum-assisted delivery.
- IVF pregnancy complicated by polyhydramnios.
- After multiple failed intubation attempts, a pediatric emergency room physician secured an airway.
- Suspected tracheoesophageal fistula prompted transfer to our institution.
- Failed tracheal reintubation after accidental extubation in NICU.
- Intentional intubation of the esophagus in operating room improved oxygen saturation.
- Angiocatheter of the stomach relieved gastric distention
- Rigid bronchoscopy revealed **what were thought to be** two cartilaginous bronchi originating from the esophagus and a posterior fistula to the stomach. ET tube secured over bronchi structures.
- Findings on CT scan interpreted as type III tracheal agenesis
- Patient transferred to a specialized quaternary care center, where further imaging revealed a type II tracheal agenesis and underwent slide tracheoplasty.



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Discussion

- Avoidance of esophageal intubation is a fundamental principle.
- Emergent esophageal intubation should be attempted only with certainty that tracheal intubation is not feasible and other more common causes of respiratory distress have been excluded.
- Case was initially misdiagnosed as tracheoesophageal fistula at an outside hospital.
- Absence of a true proximal trachea precludes use of emergency cervical exploration and tracheostomy.
- Floyd's Type III tracheal agenesis is as yet not amenable to surgical repair and is experimental.
- Luckily, subsequent esophagoscopy and imaging showing a Type II tracheal agenesis.
- A staged reconstruction was achieved at a specialized center with decompressive gastrostomy, TEF ligation, and ultimately slide tracheoplasty under cardiopulmonary bypass.

Conclusions

- While esophageal intubation is often antithetical to standard anesthesia practice, it can be a life-saving measure in the case of tracheal agenesis.
- Management of tracheal agenesis requires a high index of suspicion, prompt diagnosis, stabilization, a multidisciplinary approach, and surgical expertise for definitive correction.
- Any diagnosis with a uniformly fatal prognosis needs to be confirmed with all certainty for any goal of care discussion.

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