Tracheal agenesis: Salvaging the unsalvageable

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When Floyd and colleagues¹ described tracheal agenesis in their seminal 1962 report, there were only 12 other such cases reported in the literature. From their review of that clinical experience, the Floyd classification scheme for tracheal agenesis evolved. At the time, none of the reported patients had survived, including the subject of their report. The patients were either stillborn or died shortly after birth. Floyd type II tracheal agenesis accounts for 50% to 60% of all cases of tracheal agenesis.² In this subtype, the bronchi and bifurcation are normal, with complete atresia of the proximal airway. This type of tracheal agenesis and its surgical management is the subject of the case report in this issue of the *Journal* by Densmore and colleagues.³ Salvage treatment of a critically ill neonate with tracheal agenesis is contingent on the presence of a tracheoesophageal or bronchoesophageal fistula, which allows esophageal intubation and mechanical ventilation. Prenatal diagnosis is rare and only suspected in the *absence* of a tracheoesophageal fistula. Absence of a tracheoesophageal fistula results in congenital high airway obstruction. Congenital high airway obstruction presents with a dilated, fluid-filled, blind ending proximal tracheal pouch, absence of air in the distal bronchi, and enlarged, echogenic lung fields. In the absence of congenital high airway obstruction, the diagnosis is suspected after birth when there is absence of a cry, respiratory distress without appropriate air entry into the lungs, and failure of endotracheal intubation.⁴ Esophageal intubation is lifesaving.

Tracheal agenesis is a nearly uniformly fatal disease after birth. Surgical repair of this congenital anomaly in survivors outside of the United States has used "neotrachealization" of the esophagus with external stenting. In 4 survivors reported

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Central Message

Salvage treatment of critically ill neonates with tracheal agenesis requires innovative surgical strategies, dedicated resources, and the coordinated effort of multiple medical specialists.

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from Osaka University, gastrointestinal continuity was established by gastric or jejunal interposition.⁵ Densmore and colleagues³ have modified this surgical approach to save a particularly high-risk patient who had a nearly atretic tracheoesophageal fistula and concerns for neurologic insult from postnatal hypoxia. Their report speaks to the coordinated effort required of multiple medical specialists, and the resources required at a tertiary care facility, to achieve a truly remarkable outcome for the patient and family.

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