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LETTER

Management of congenital tracheal agenesis

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Heimann et al. [2] reported three patients with agenesis of the trachea. Although a very rare disease, tracheal agenesis is of eminent importance for neonatologists when they are confronted in the delivery room with a hypoxic neonate and the inability to perform endotracheal intubation. So they are challenged not only to achieve adequate oxygenation but also to quickly confirm whether the patient has a lethal or potentially curable malformation of the respiratory system.

Extending the data of Heimann et al. we report two additional patients with tracheal agenesis. Both presented postnatally with cyanosis and the absence of audible cry. Although endotracheal intubation failed, adequate oxygenation was achieved via transoesophageal ventilation since a broncho-oesophageal fistula was patent in our patients as well as in almost all of those previously described.

Ventilation through a tube, inserted orally into the oesophagus, might be the only option to gain time for further investigations. The diagnostic tool of choice is laryngobronchoscopy combined with a computed tomography (CT) scan or magnetic resonance imaging. Rapid diagnosis is vital to differentiate between a high obstructive airway syndrome (HOAS) like laryngeal atresia, in which tracheotomy would be life-saving, and tracheal agenesis, where potential surgical options depend on the extent of atresia present [3]. As in our first patient laryngobronchoscopy or CT scan were not available in the delivering hospital, explorative surgery with incision of the upper thoracic aperture was performed under transoesophageal ventilation and tracheal agenesis was confirmed. Transoesophageal ventilation achieved oxygenation; however, hyperinflation of the stomach occurred because of an imperforate anus. Gastrostomy was performed to decompress the gastrointestinal tract. A Fogarty catheter was inserted into the stomach, placed into the distal part of the oesophagus with the intention to block the distal oesophagus; but after incision, the opened stomach presented as a major 'air leak' and oxygenation failed. Resuscitation was terminated 4 h after birth. Autopsy revealed Floyd type II tracheal atresia and anal atresia.

In our second patient, total tracheal agenesis was confirmed by bronchoscopy and CT. After 48 h of uneventful transoesophageal ventilation with sufficient oxygenation, ventilation was discontinued because parental consent for further surgery (banding of the lower oesophagus, insertion of gastrotube as first step) could not be obtained. The patient died 49 h after birth.

Most reported newborns with tracheal agenesis expire postnatally and long-term survival is rare [1]. However, if tracheal agenesis is considered during resuscitation, intubation of the oesophagus and ventilation via a broncho-oesophageal fistula is an option to manage these patients until an exact diagnosis can be made. Our experience emphasizes the importance of an early and rapid discrimination between tracheal agenesis and HOAS.

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