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Review Article

Clinical presentation and airway management of tracheal atresia: A systematic review

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ABSTRACT

Objectives: Tracheal atresia (TA) is a rare congenital condition that typically requires an unexpected and emergent resuscitation in the delivery room. The mortality rate associated is very high, with only a few long-term survival cases reported. We describe the findings of a systematic review on the clinical presentation and airway management of TA.

Methods: Using the keywords "tracheal atresia", "tracheal agenesis" and "tracheal hypoplasia" a search through Embase and Pubmed databases was performed following the Preferred Reporting Items for Systematic Reviews and Meta-Analyses (PRISMA) protocol. Articles published from 1950 to 2015 in English, French, Italian, Portuguese and Spanish were included. Exclusion criteria were cases of stillborn, and unclear diagnosis or outcome.

Results: 149 cases of TA were identified after reviewing 1125 initial references. There was a male preponderance (65%), and associated malformations were described in 94.2% of patients. Prenatal ultrasound was abnormal in 56.3% of cases, with polyhydramnios being the most common finding. The most frequent type of TA was Faro Type C. 94 (41.3%) patients did not survive beyond the first 24 h of life. Only 13 (8.4%) patients survived more than three months of life, after undergoing a variety of surgical approaches.

Conclusion: This review, which to our knowledge is the largest one to date, confirms that TA is a rare malformation, occurs more frequently in males, and has a very high mortality rate. Depending on the presence and type of concomitant malformation, as well of the length of the remaining trachea, different surgical management options are described.

Contents

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1. Introduction

Tracheal atresia (TA) is a rare congenital defect that is usually incompatible with life. It has an estimated incidence of 1 per 50,000 newborns and affects more males than females [1]. This defect should be suspected in any newborn presenting with severe respiratory distress and cyanosis shortly after birth, absence of audible cry, as well as failure of endotracheal intubation [2].

The diagnosis is often made in post-mortem examination, where a total or partial absence of the trachea below the larynx is revealed. A concomitant tracheo- or broncho-esophageal fistula is often seen, and its presence allows for resuscitation and ventilation through an esophageal intubation [3]. A few classification systems are suggested in the literature, with the Faro's classification being the most comprehensive [4] (Fig. 1). Survival after delivery is dependent on a prompt diagnosis, as well as proper management of the airway. Other congenital malformations are frequently present, especially in the cardiac, gastrointestinal, and genitourinary systems [5].

Since the first reported case of this malformation described by Payne in 1900 [6], over a 150 case reports of TA have been published in the literature. Only a few cases of neonates with TA have undergone successful management [7,8]. Given the low frequency of TA cases and its high-stakes outcomes, a systematic review would provide insight to physicians on when to anticipate this malformation and how to promptly diagnose and manage it. To our knowledge, we present the first systematic review of all the TA cases reported in the literature, where we discuss the varied clinical presentation of this anomaly and discuss the emergent and surgical airway management options.

2. Material and methods

Institutional review board approval and patient consent were not required due to the nature of this study. A comprehensive search for all studies published up to October 2015 was performed using PubMed and Embase, which included MEDLINE and other Elsevier journals. One author conducted the search with the

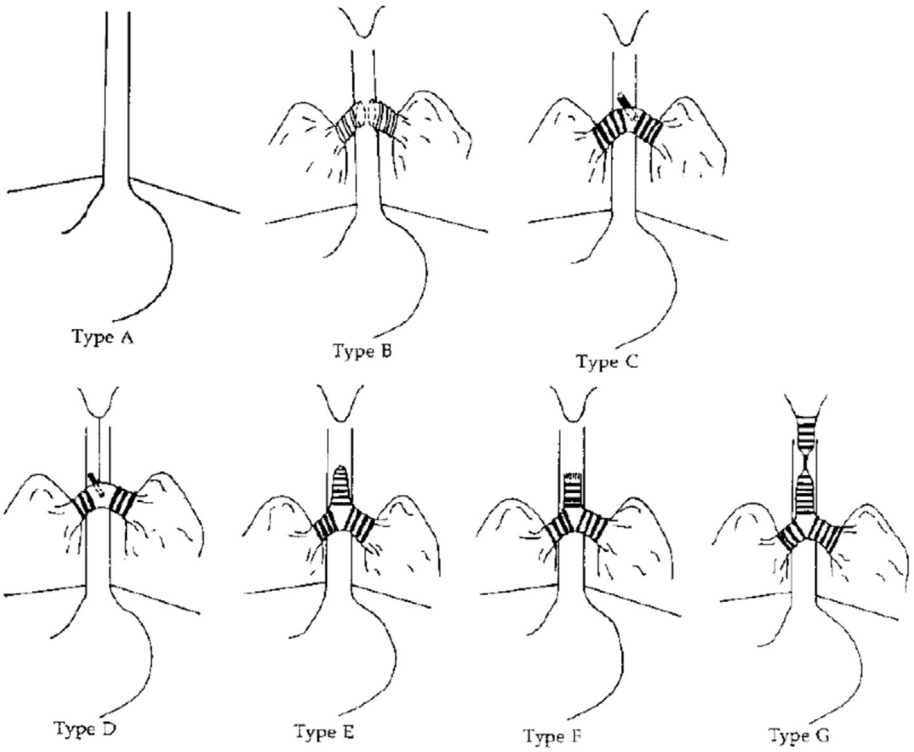

Fig. 1. Faro's Classification System (borrowed with permission from Saleeby et al. [9]).