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Esophageal duplication and congenital esophageal stenosis

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ABSTRACT

Esophageal duplication and congenital esophageal stenosis (CES) may represent diseases with common embryologic etiologies, namely, faulty tracheoesophageal separation and differentiation. Here, we will re-enforce definitions for these diseases as well as review their embryology, diagnosis, and treatment.

Introduction

Congenital foregut malformations present as a wide variety of surgically important entities. Of these, congenital malformations of the esophagus are particularly relevant to pediatric surgical practice. This article examines two of these disease processes in detail: esophageal duplication and congenital esophageal stenosis (CES).

While often discussed as distinct clinical entities, esophageal duplication and CES may have similar but as of yet incompletely understood embryologic origins.[1] Both are products of abnormal foregut embryogenesis and may represent points along a spectrum of disease that include esophageal atresia with or without tracheoesophageal fistula (EA/TEF) as well as congenital bronchopulmonary malformations.[2] There has historically been some confusion regarding the definitions of esophageal duplication and congenital esophageal stenosis in the literature due to the wide range of diagnoses, common origins, and incomplete understanding of embryogenesis.

The term, "esophageal duplication," as it is commonly used, describes three different morphologic variants of the more inclusive term, "foregut duplication." The morphologic variants are as follows: (1) cystic, (2) tubular, and (3) diverticular.[3] Classically, esophageal duplication had to have the following three characteristics: (1) a well-developed coat of smooth muscle, (2) an epithelial lining representing some portion of the alimentary tract (though some respiratory components may be present), and (3) an attachment to the esophagus.[4] We prefer this classical definition but realize that some authors have used the term, "esophageal duplication" to describe any thoracic duplication containing purely gastrointestinal epithelium even if the duplication is remote from the esophagus.[4] When containing respiratory epithelium and having close anatomic relation to the trachea or bronchi, foregut duplication cysts are more aptly named, "bronchogenic cysts".[1] "Neurenteric cysts" are another important subset of foregut duplication and represent foregut duplications that have extension into the spinal canal. These should be recognized as a distinct clinical entity given the complexity of their disease associations, diagnosis, and treatment.[5–7] We will limit our discussion here to esophageal duplication as defined above.

CES as discussed here will represent three distinct lesions: (1) tracheobronchial remnants, (2) fibromuscular stenosis, and (3) membranous webs.[8] The stenosis in these cases represents a congenital narrowing due to a mural process rather than from external compression from mass effect, hence their inclusion under this umbrella term though they may prove to have different etiologies in the future. These must also be distinguished from causes of acquired esophageal stenosis, primarily strictures from gastroesophageal reflux.

Embryology

From a conceptual standpoint, the esophagus' function is simple; its only purpose is antegrade propulsion of liquid and masticated food from the mouth to the stomach. This simplicity belies the complexity of foregut development. The separation of the primitive foregut tube into the esophagus and tracheobronchial tree is key to the normal development of the esophagus and respiratory system. Aberrations in normal development can lead to a wide variety of disease states. The mechanisms by which the

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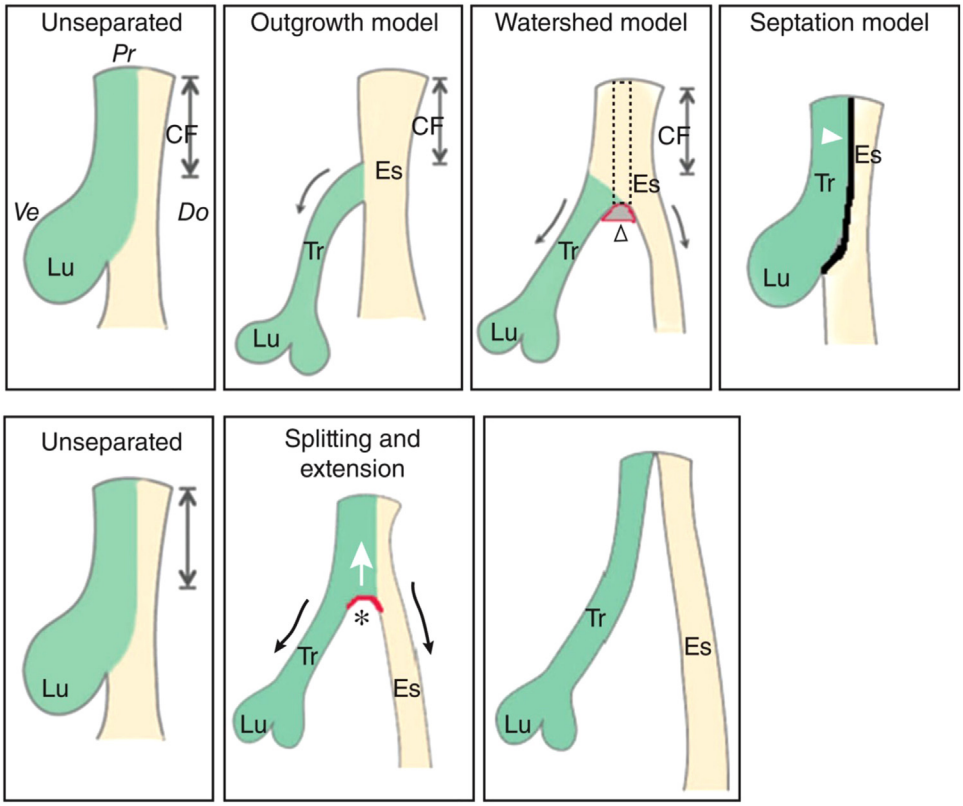

Fig. 1. Models of tracheal-esophageal separation: (1) The outgrowth model—trachea extends from the common foregut tube as the lung buds grow, while common foregut tube becomes the esophagus. (2) The watershed model—both the trachea and esophagus elongate while separated by a mesenchymal septum that serves as a wedge to prevent the extension of the lateral wall at the dorsal-ventral midline. The empty arrowhead represents the hypothetical mesenchymal condensation, which has yet to be identified. (3) The septation model—epithelial cells at the dorsal-ventral midline make contact across the lumen and fuse to form a septum. (4) Splitting and extension—proposes that the separation of the trachea and esophagus initiates at the level where the lung grows out and moves rostrally. A saddle-like structure (red arc) splits the anterior foregut. Abbreviations: Ve, ventral; Do, dorsal; Pr, proximal (rostral); Di, distal (caudal); CF, common foregut; Lu, lung; St, stomach; Es, esophagus; Tr, trachea. (Adapted with permission from Que J.[16]) (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)

endodermal-derived foregut tube separates into the esophagus and trachea are still being elucidated. No consensus exists regarding not only the morphological changes that occur during this critical period but also regarding the complex array of cell signaling interactions that drive them.[1,9,10]

What is clear is as follows: tracheoesophageal separation occurs between the fourth and fifth week of life.[11] Many mechanisms have been proposed regarding this process. The origin of the so-called tracheoesophageal septum continues to be unclear. Historical descriptions of key structures are credited to His in 1887 and Rosenthal in 1931.[12–14] In these classic descriptions, endodermal ridges form in the lateral walls of the primitive foregut, fusing in a caudal to cranial direction like a "zipper" separating the primitive esophagus and trachea.[1] Three contemporary models have been proposed, the first two as a challenge to the latter and the third in an attempt to make it more robust[15,16] (Figure 1). The first contemporary theory, the outgrowth model, directly challenges the appearance of lateral ridges and instead proposes budding and growth of the trachea outward from the common foregut tube, which becomes the esophagus.[17,18] The second contemporary theory, the watershed model, proposes that a mesenchymal wedge exists to prevent growth and extension of the lateral walls of the common foregut tube while the trachea and esophagus/pharynx elongate.[19] The third model, the septation model, builds upon the classical description mentioned above, proposing that lateral edges of the primitive foregut fuse to form the tracheal and esophagel compartments.[20] Lack of experimental evidence and correlation with disease states is commonly cited as the reason any one of these models is not favored over the others.[15,16] In the septation model, the lack of evidence by imaging (electron microscopy of chick embryos) of developing lateral folds is at odds with this model's accuracy. However, Que[16] and Metzger et al.[10] have recently described a saddle-like structure that has been experimentally observed and is supportive of a variation of the septation model. Que has deemed this new model, the "splitting and extension model."

The above models describe morphogenic behavior irrespective of the molecular signaling occurring at the point of differentiation. Much progress has been made in the characterization of these signals (Figure 2). It is clear that reciprocal signaling occurs between the endodermal foregut derivative and surrounding structures, notably the mesenchyme and notochord. The

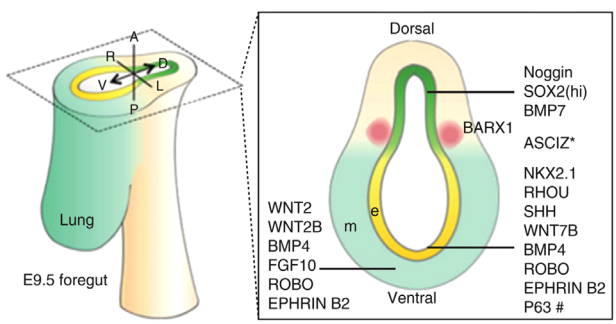

Fig. 2. Dorsal-ventral pattern of signaling during tracheal-esophageal separation. (Adapted with permission from Que J.[16]) (For interpretation of the references to color in this figure legend, the reader is referred to the web version of this article.)