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Congenital tracheobronchial stenosis

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ABSTRACT

Congenital tracheobronchial stenosis is a rare disease characterized by complete tracheal rings that can affect variable lengths of the tracheobronchial tree. It causes high levels of morbidity and mortality both due to the stenosis itself and to the high incidence of other associated congenital malformations. Successful management of this complex condition requires a highly individualized approach delivered by an experienced multidisciplinary team, which is best delivered within centralized units with the necessary diverse expertise. In such settings, surgical correction by slide tracheoplasty has become increasingly successful over the past 2 decades such that long-term survival now exceeds 88%, with normalization of quality of life scores for patients with non-syndrome-associated congenital tracheal stenosis. Careful assessment and planning of treatment strategies is of paramount importance for both successful management and the provision of patients and carers with accurate and realistic treatment counseling.

Introduction

Congenital tracheal stenosis (CTS) is a rare disease estimated to affect 1 in 64,500 live births characterized by complete tracheal rings, localized to any part of the large airway. It is typically classified according to the length of the affected area as either short segment or long segment.[1] At birth the tracheal length is approximately 3 cm. Lesions are generally considered long segment when over 1 cm in the newborn and 1.5 cm in the infant, or classified according to the percentage of trachea affected such that $>50\%$ is considered long-segment disease.[2,3]

A total of 60% of children born with CTS often have other associated malformations, particularly cardiovascular anomalies. It is the combination of both airway and cardiovascular disease that often leads to life-threatening compromise.[2] Other congenital malformations include gastrointestinal malformations and anorectal anomalies. Due to its significant associated morbidity and mortality, the precise diagnosis and anatomical delineation of CTS must be made to ensure that not only an appropriate treatment strategy is chosen but also realistic and accurate pretreatment counseling of parents/carers can be given. The rarity of this disease also mean that the care centers that can manage this condition are limited to fewer tertiary units with the necessary team-based expertise required to manage a child with this complex condition.

Treatment strategies for children with CTS have evolved over the past 2 decades such that life expectancy has significantly improved.[4] Original strategies are focused upon end-to-end resection and primary anastomosis: however, due to tensions placed on the anastomosis, such an approach was only possible on short-segment disease. Alternative methods were required for long-segment congenital tracheal stenosis (LSCTS), and many clinicians attempted anterior augmentation with an anterior patch (pericardial, cartilage, etc.) combined with suspension sutures or stents.[5–7] While this surgical technique would overcome the problems of the immediate stenosis, long-term results were often complicated by persistent granulation and restenosis.[8,9] Other strategies included tracheal replacement with aortic and tracheal homografts[10–12], but these were similarly associated with significant morbidity and variable outcomes. Slide tracheoplasty was first used for CTS in 1989[13] and has subsequently proved to be extremely versatile; it has, therefore, been adopted by many units as the surgical treatment for LSCTS. Its use has been extended to both acquired lesions of the trachea such as iatrogenic stenosis and short-segment disease, with large series confirming it to be a safe and reliable technique with survival rates of over 88% and low associated morbidity and mortality.[14–16]

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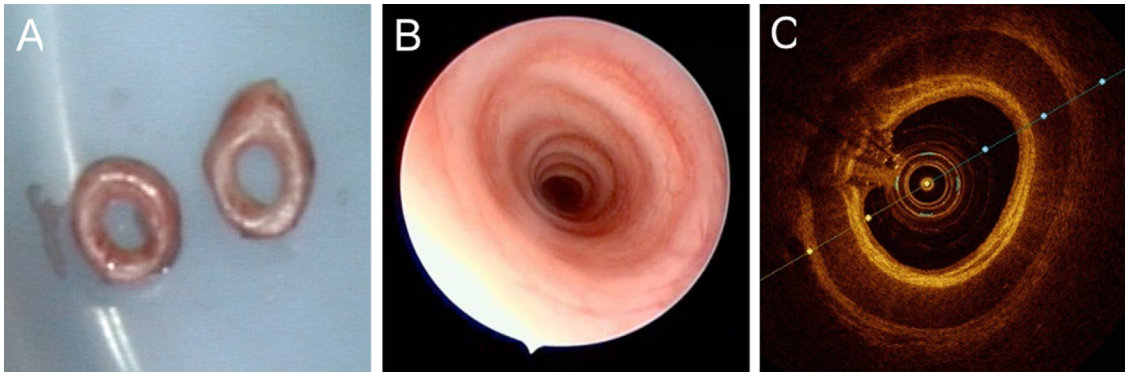

Fig. 1 (A) Macroscopic images of complete tracheal rings found in LSCTS following resection. (B) Endoscopic appearance of congenital tracheal stenosis demonstrating the luminal appearance of complete rings. (C) Optical coherence tomography (OCT) imaging demonstrating the appearance of full "O shape" rings and intact cartilage.

Embryology

An understanding of aerodigestive tract allows an appreciation of the myriad of associated congenital anomalies seen. At the 3rd week, the laryngotracheal groove or sulcus appears in the proximal foregut and progresses caudally alongside the cephalad progression of the lateral ridges, thereby forming the primordial trachea. The bronchial primordia develop soon after asymmetric tracheal tip buds and the pulmonary primordia as ventral bulges of the foregut. Complete separation of the trachea and esophagus has usually occurred by the 6th week with the tracheal bifurcation descending gradually to the level of the 4th vertebra. Cartilage, muscle, and connective components are derived from proliferation of coelomic cavity cells, and cartilage is noted from the 10th week. Concurrent to this, the glottis forms from a median slit in the pharyngeal floor between the 4th and 6th branchial arches with a "T" shape opening into a lumen at around the 8th week. By this time the thyroid and cricoid cartilaginous framework has appeared, derived from the 4th and 6th arches, followed by vocal cords at the 12th week.

Anomalies can occur at any point along the tract, the most common being failures in separation of the airway and the alimentary tracts resulting in tracheoesophageal fistulas distally and laryngotracheoesophageal clefts superiorly. Segmental failure leads to atresia, and in some cases, complete agenesis that is typically fatal. The relatively separate development of the laryngeal, trachea/esophagus, bronchial, and pulmonary complexes allows a diverse configuration of anomalies that may include agenesis and/or stenosis of the aerodigestive tract, either as isolated segments or as combinations of these 4 components.[17]

Classification and anatomical variants

Normal tracheal stenosis is predominately caused by developmental anomalies in the cartilaginous exoskeleton. Occasionally, a short stenosis can be caused by an absent cartilaginous section typically limited to 1 or 2 rings. The most common finding is a segment of complete cartilaginous rings with a luminal diameter as small as 1–2 mm.[2] The normal tracheal and bronchial cartilages are "C" shaped with a membranous tracheal muscle running posteriorly. They are suspended from the cricoid, the only complete ring of cartilage in the normal airway, which is pivotal due to its role in opening and closing the glottis through its articulation with the thyroid cartilage. This complete cricoid ring is susceptible to acquired stenosis from the trauma of intubation with an endotracheal tube (ET). Children with LSCTS can, therefore, present with an acute airway crisis due to ET traumatization of the upper end of the stenosis that causes further airway narrowing via inflammation, granulation, and scarring.

CTS is predominately caused by developmental anomalies in the cartilaginous exoskeleton. The most common finding is a segment of complete cartilaginous rings (Figure 1) with a luminal diameter as small as 1–2 mm,[2] though an absent cartilaginous section can occasionally cause a short stenosis typically limited to one or two rings. The pattern, severity, and extent of the complete cartilaginous rings causing the stenosis are variables, occurring in multiple configurations throughout the tracheobronchial tree. A variety of classification systems have been proposed based upon disease extent and severity; however, no consensus on classification has been reached due to the heterogeneous nature of the condition. In our unit, arborization patterns have been classified as normal bifurcation, anomalous right upper lobe or porcine bronchus (bronchus suis), bronchial trifurcation, or as single lung.[18,19] (Figure 2) This is further classified into congenital tracheal rings limited to the trachea only or involving the bronchi and is known as congenital tracheobronchial stenosis (CTBS). In some cases, a bronchial stenosis with a few rings affected may be the only finding. As expected from the embryological development of the airway, other structural anomalies may also be found including subglottic stenosis, tracheoesophageal fistula, and esophageal atresia.

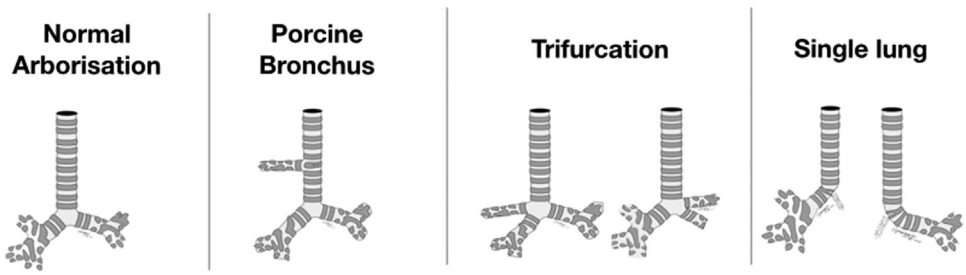

Fig. 2. Typical arborization patterns found in congenital tracheal stenosis. These include a normal airway, pig bronchus, trifurcation, and single lung configurations. Congenital tracheal stenosis can be limited to the trachea alone or extend into any 1st division bronchus.