Seminars in Pediatric Surgery 25 (2016) 156–164

Contents lists available at ScienceDirect

Seminars in Pediatric Surgery

journal homepage: www.elsevier.com/locate/sempedsurg

Pediatric tracheomalacia

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ARTICLE INFO

Keywords:
Tracheomalacia
Tracheobronchomalacia
Aortopexy
Tracheobronchopexy
External airway fixation
Bronchoscopy

ABSTRACT

Tracheomalacia (TM) is defined as an increased collapsibility of the trachea due to structural anomalies of the tracheal cartilage and/or posterior membrane. Tracheomalacia has a wide range of etiologies but is most commonly present in children born with esophageal atresia and tracheal esophageal fistula. Clinical symptoms can range from minor expiratory stridor with typical barking cough to severe respiratory distress episodes to acute life-threatening events (ALTE). Although the majority of children have mild-to-moderate symptoms and will not need surgical intervention, some will need life-changing surgical treatment. This article examines the published pediatric literature on TM, discusses the details of clinical presentation, evaluation, diagnosis, and a variety of treatments.

Introduction

Tracheomalacia (TM) refers to a weakness of the trachea, frequently due to reduction and/or atrophy of the longitudinal elastic fibers of the pars membranacea, or impaired cartilage integrity, such that the airway is softer and more susceptible to collapse with changes in pressure.[1] In 1963, TM was described by Baxter and Dunbar as an uncommon condition in which the tracheal wall is especially soft and pliable.[2] This lack of stiffness results in an abnormal occlusion of the tracheal lumen.[3] The process can involve the whole trachea or can be localized to one segment only.[4] If the mainstem bronchi are involved as well, the term tracheobronchomalacia (TBM) is employed. The term bronchomalacia (BM) is used to describe the isolated weakness and collapsibility of one or both of the mainstem bronchi without tracheal involvement. It is much less common than TM or TBM.[1]

During normal inspiration, the intrathoracic tracheal lumen dilates, and during expiration it narrows because the airway diameter is in part determined by the difference between intrathoracic and intraluminal airway pressures.[5] Even when very high intrathoracic pressures are generated, the rigidity of the cartilage in the tracheal wall prevents complete airway collapse.[6] In patients with TM, the abnormal collapsibility of the trachea accentuates the physiological airway narrowing that occurs during expiration, and in severe cases clinically obvious obstruction of the airway may result when greater intrathoracic pressure increases occur, as during forced expiration or coughing.[7] In the extreme case, complete obstruction to the outflow of air may lead to a fatal outcome.[8] In addition, the lack of normal tracheal stiffness in TM allows tracheal collapse from compression by adjacent thoracic structures, mainly the aortic arch and innominate artery anteriorly and the esophagus posteriorly.[9] The aortic size and position are relatively constant, although subtle changes in position and anatomy can have profound effects on the developing airway. The size of the esophagus increases with swallowing, gastroesophageal reflux, and in the presence of stenotic or obstructive lesions of the lower esophagus,[10] which may compress the trachea posteriorly. Therefore, patients with TM often have their most severe symptoms during or shortly after eating.[11]

Classification

TM and TBM have been classified in many ways, but we adhere to the more commonly used division into congenital or primary and acquired or secondary[1] (Table 1).

Congenital TM was described by Holinger et al.[12] and later defined as weakening of the tracheal wall due to abnormal formation on maturation of the airway.[13] The incidence of congenital TM is approximately 1:2,100 children, and it is the most common congenital tracheal abnormality.[14,15]

TM and TBM are common respiratory problems in children with esophageal atresia (EA).[16] The prevalence of severe TM among EA has been reported to be 11–33%,[17,18] and TM/TBM are

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http://dx.doi.org/10.1053/j.sempedsurg.2016.02.008
1055-8586/

Table 1
Diseases associated with TM-TBM (modified from Carden KA et al (1).

Primary/Congenital TM-TBM	Secondary/Acquired TM-TBM
Idiopathic TM – normal infant	Prolonged intubation
Prematurity	Tracheostomy
Cartilage congenital abnormalities	Severe tracheobronchitis
– Dyschondroplasia/chondromalacia/ chondrodysplasia	Resulting from compression
– Polychondritis	Vascular
– Ehlers-Danlos syndrome	– Double aortic arch
Congenital syndromes	– Abnormal take-off the innominate artery
– Mucopolysaccharidosis (Hunter and Hurler)	– Right aortic arch
– CHARGE* syndrome/VATER association**	– Aberrant right subclavian
– Trisomy 9 and 21	– Enlarged pulmonary veins
– Antley-Bixler	Cardiac
– Hallermann-Streiff	– Left atrial hypertrophy
– Crouzon	– Enlarged left atrium
– Pfeiffer	Skeletal
– Blackfan-Diamond	– Scoliosis
– Williams-Campbell	– Pectus excavatum
– DiGeorge	Tumors and cysts
– Larsen and Larsen-like	– Teratomas
– Brachmann de Lange	– Bronchogenic cyst
– Robin Sequence	– Enterogenic cysts
– Atelosteogenesis type 1	– Thymomas
– Deletion 11p13, 22q11 and 12q	– Lymphatic malformation
– Translocation 18-22	– Lymphoma
– Kniest dysplasia	– Neuroblastoma
– Camptomelic dysplasia	– Cystic hygromas
Congenital anomalies	– Hemangiomas
– Tracheoesophageal fistula	Infection
– Esophageal Atresia with or without laryngeal cleft	– Abscess
– Bronchopulmonar dysplasia	Posttraumatic

* CHARGE = colobomata of the eyes, heart defects, choanal atresia, retardation of growth, genital hypoplasia and ear abnormalities.
** VATER = vertebral defect, anal atresia, tracheoesophageal fistula, esophageal atresia and radial/renal dysplasia.

more frequent in association with EA with tracheal esophageal fistula (TEF) than with pure EA.[19] The airway collapse may be attributable to congenital weakness and malformation of the cartilage rings causing anterior collapse, and with increase in the length of the posterior membranous muscle contributing to posterior airway intrusion.[20] In 1979, Wailoo and Emery[21] conducted postmortem examinations on the tracheas of 40 children born with EA and TEF who had died before treatment. They described an elliptical deformity (flattening) of the tracheal lumen, deficiency of cartilaginous rings with a concomitant increase in the length of membranous muscle.[21] There was a decrease in the normal 4.5:1 circumference ratio of cartilage to membranous trachea in most of these children.[22] This resulted in a loss of normal horseshoe (or "C") shape of the cartilages into more "U" shaped or even broad-based "Bow" shaped cartilages, resulting in progressively wider posterior membranes potentiating the posterior membrane intrusion into the tracheal lumen, which is interpreted as anteroposterior collapse of the wall during the exhalation phase of respiration[1,21] for the intrathoracic trachea. These pathological findings correspond precisely to those seen at bronchoscopy in children with symptomatic TM and EA-TEF (Figure 1).

Two hypotheses have been postulated to explain the association between TM and EA-TEF. Faulty division of the primitive foregut was proposed by Emery and Haddadin,[23] who observed the inclusion of esophageal muscle and squamous epithelium in the membranous part of the trachea in about 80% of the children with EA but only 2% of the normal babies. The abnormalities seen at autopsy by Wailoo and Emery[21] in children with TM and EA lend additional support to this supposition. Davies and Cywes[24] suggested that dilatation of the proximal esophageal pouch compressed the trachea *in utero* and altered its normal development. In addition, Wailoo and Emery suggested that the loss of normal intratracheal pressure *in utero* through a TEF might lead to a more flaccid trachea.[25]

While congenital TM can occur alone, it is usually associated with other developmental airway anomalies such as laryngomalacia, bronchomalacia, and laryngeal clefts.[26,27] It can additionally be seen in conjunction with several syndromes, chromosomal defects, mucopolysaccharidose deficiencies, and connective tissue disorders [28] (Table 1).

Acquired or secondary TM is more common than the congenital form, and it is caused by the degeneration of normal cartilaginous support from a variety of causes.[2] Secondary TM is also associated with other thoracic lesions such as vascular rings and tumors,[11] although this may be thought of as cartilaginous deformation from external pressure resulting in airway stenosis without significant dynamic collapse. Cartilaginous defects causing TM can be acquired from infection[29,30] and long-term ventilation with high airway pressure (Table 1).[31]

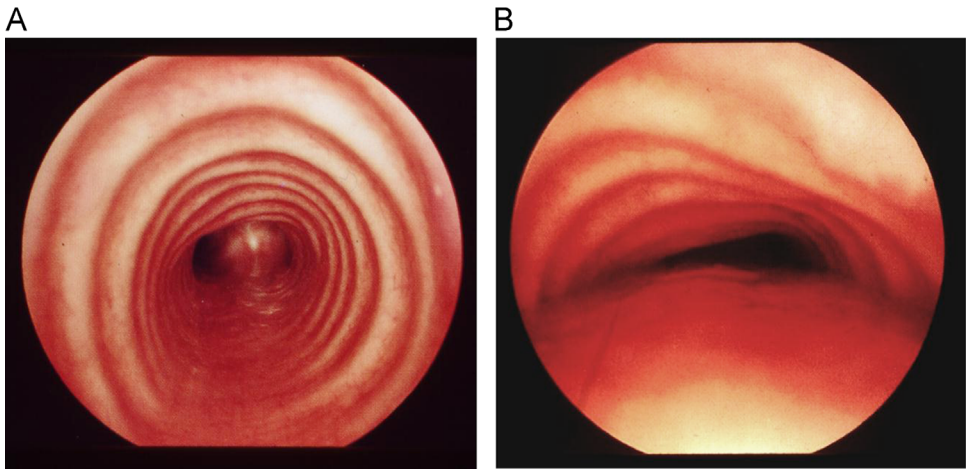

Fig. 1. (A) Endoscopy of normal trachea—proportion between cartilage to membranous trachea is about 4.5:1 ratio and (B) endoscopy of newborn after surgery for correction of esophageal atresia and distal tracheoesophageal fistula. This newborn could not be extubated after surgery. Documented severe TM with almost complete tracheal collapse with exhalation, and the proportion of cartilage to membranous trachea is much less than 4.5:1 ratio.