Tracheomalacia

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KEYWORDS

• Tracheomalacia • Airway collapse • Dynamic airway CT • Aortopexy • Tracheoplasty

KEY POINTS

- Tracheomalacia is characterized by excessive collapsibility of the trachea, typically during expiration.
- Dynamic airway CT as a modification of the standard protocol seems to be a promising tool for noninvasive diagnosis of airway malacia.
- Surgery should be considered in severe symptomatic disease, especially in localized and segmental forms. Surgical lateropexia, tracheal resection, and surgical external stabilization are options.
- Tracheoplasty seems to be the best choice for selected cases of adult malacia.
- Surgery in children is different, due to the different causes. The most commonly performed surgical method is aortopexy.

TRACHEOMALACIA IN GENERAL

Malacia of the trachea, or tracheomalacia, comprises different conditions of the trachea that have a common impact: the tracheal walls are in close proximity. However, this manifests in different ways and from different causes. Tracheomalacia is both a dynamic and a fixed state. In general, the trachea changes its shape physiologically during the breathing cycle, depending on the pressure impact. Normally, there are certain phases during which the tracheal walls converge. The dynamic form of tracheomalacia may lead to an almost complete airway obstruction during regular breathing because the tracheal lumen is reduced to a slit or it is impassable (**Fig. 1**). This effect is significantly enhanced by extreme breathing maneuvers, such as coughing or the Valsalva maneuver. Tracheomalacia can affect the trachea in its complete length, only in certain segments, or in multiple segments. In addition, tracheomalacia can extend to the mainstem bronchi (tracheobronchomalacia).

Tracheomalacia is generally caused by one of two mechanisms: altered structures of the tracheal wall (eg, alterations of the tissue architecture or the elastic tissue characteristics)[1,2] or acquired secondary factors (eg, external compression or chronic inflammatory response).[3]

CLASSIFICATION OF TRACHEOMALACIA

In the literature, there are different approaches that systematically classify tracheomalacia based on different perspectives.[4] It seems rational to differentiate between tracheomalacia in children and adults.[5] Approaches that considered the different forms of the trachea lead to the classifications of saber-sheath (**Fig. 2**) and crescent shape.[6]

In 2005, Carden and colleagues[7] proposed a classification for tracheomalacia in both children and adults based on comprehensive reviews that differentiated between primary congenital causes and secondary acquired causes (**Boxes 1** and **2**).

In general, the well-known classification systems for tracheomalacia are organized merely descriptively and by factors from which no systematic considerations for treatment can be derived. In 2007, Freitag and colleagues[8] created

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Thorac Surg Clin 24 (2014) 51–58
http://dx.doi.org/10.1016/j.thorsurg.2013.09.003

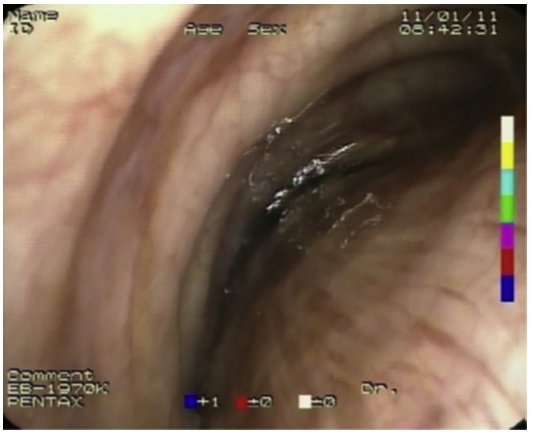

Fig. 1. Expiratory tracheal collapse in tracheomalacia due to chronic bronchitis.

a system for comparing therapeutic decisions and methods.

DIAGNOSIS OF AIRWAY MALACIA

Patients with airway malacia typically present with nonspecific chronic respiratory complaints, including dyspnea, cough, and recurrent infections. Thus, it is not possible to make the diagnosis of malacia based on clinical symptoms alone. Pulmonary function testing may provide supportive evidence of airway malacia but it is not diagnostic. Typically, there is a rapid decline of expiratory flow after a sharp peak that is associated with the collapse of central airways due to negative transmural pressure in the flow-volume curves. Conversely, the volume curve may be typically cropped due to stenosis. Despite sometimes typical findings, estimation of the severity cannot be made by pulmonary lung function testing.

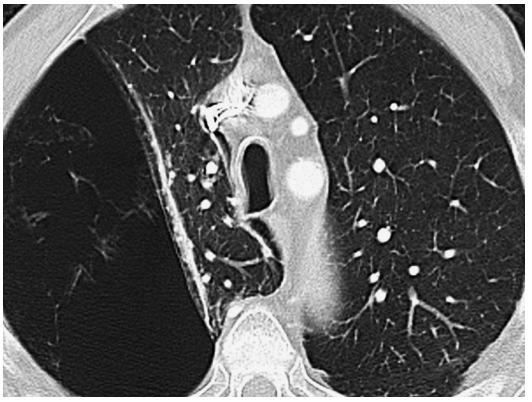

Fig. 2. Typical finding of a saber-sheath trachea. In this case, the lateral walls converge instead of the anterior and posterior tracheal walls.

Box 1
Classification of tracheomalacia in children

Congenital:
- Idiopathic
- Prematurity
- Congenital abnormalities of the cartilage
- Congenital syndromes associated with tracheomalacia
- Congenital anomalies associated with tracheomalacia

Acquired:
- Prolonged intubation
- Tracheotomy
- Severe tracheobronchitis
- External compression
  - Vascular
  - Cardiac
  - Skeletal
  - Tumors and cysts
- Infection
- Posttraumatic

Data from Carden KA, Boiselle PM, Waltz DA, et al. Tracheomalacia and tracheobronchomalacia in children and adults: an in-depth review. Chest 2005; 127:984–1005.

However, central airway collapse is not correlated with the degree of obstruction as assessed by forced expiratory volume in 1 second and central airway collapse may be found irrespective of the degree of expiratory flow limitation during quiet breathing.[9] There is no significant correlation between expiratory tracheal collapse and any pulmonary function measure. In a recent study, excessive expiratory tracheal collapse was observed in a subset of subjects with chronic obstructive pulmonary disease (COPD). However, the magnitude of collapse was independent of disease severity and did not correlate significantly with physiologic parameters.[10] There is a need for a standardized approach to evaluate airway malacia objectively in the patient with dyspnea refractory to traditional therapies.[11]

When pediatric pulmonologists expected to find airway malacia (based on symptoms, history, and lung function) before bronchoscopy, they were correct in 74% of the cases. In 52% of the airway malacia diagnoses, the diagnosis was not suspected before bronchoscopy.[12] Congenital malacia of the large airways is one of the few causes