



Epidemiology and Management of Pediatric Tracheomalacia

**Haya Jamal M. Albalawi^{1*}, Rawan Defallah G. Alzahrani¹,
Rana Suliman D. Alshwamin¹, Raghad Turki Asiri¹,
Sarah Awad Muhammad Alenzi², Amal Abutaleb Qaysi²,
ALanoud Abdullah H. Althobaiti², Nashwa Zead A. Albalawi²,
Raghad Fraih A. Albalawi², Rahaf Saad Alharbi³
and Amal Mohammad H Albalawi⁴**

¹Medical Intern at University of Tabuk, Saudi Arabia.

²University of Tabuk, Saudi Arabia.

³University of Taibah, Saudi Arabia.

⁴Vision College, Saudi Arabia.

Authors' contributions

This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.

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ABSTRACT

Tracheomalacia is excessive trachea collapse, usually upon expiration. Severe symptoms accompany congenital types. Often milder versions occur following the neonatal period. Adult malacia is mostly linked to pulmonary chronic obstruction. There's still no standardized functional bronchoscopy. Dynamic CT airway is an excellent diagnostic tool. Bronchoscopy and stent insertion led to a major improvement, although with a high incidence of complication. Surgical lateropexia, excision of trachea and external surgical stabilization are possibilities. Aortopexy is the most common operation in children.

Keywords: Etiology; tracheomalacia; laryngomalacia; bronchopulmonary.

**Corresponding author: E-mail: hayaalbalawi9@gmail.com;*

1. INTRODUCTION

Tracheomalacia is a condition in which the tracheal walls collapse abnormally. It can be detected as an individual lesion or in combination with other lesions that produce airway tightness or injury. It is a benign condition characterized by symptoms caused by airway blockage. As a result, this condition is frequently misdiagnosed as chronic asthma or as acute inflammatory injury of the bronchioles [1] although there is no universally accepted classification system, the disease's etiology can be divided into three categories: congenital tracheal abnormalities, acquired disorders weakening the whole or partial tracheal structure, and conditions causing extrinsic airway compression. [2] The weakening of several cartilaginous arches, especially in the distal portion of the trachea, is common in primary tracheomalacia. Acquired tracheomalacia is frequently caused by widespread infection and inflammation, as observed most commonly after a case of tracheitis or any other viral or bacterial infection [3].

With an estimated frequency of at least 1 in 2,100 children, primary airway malacia is not common in the general population. Clinical symptoms of airway malacia overlap with those of more prevalent lung disorders, making it difficult to diagnose [4]. Depending on Patients' symptoms tracheomalacia may spontaneously resolve over the natural history of the disease or can cause persistent respiratory distress [2, 5-7].

1.1 Etiology

Tracheomalacia can be broadly considered as being congenital or acquired [8], however children's tracheal pathology is almost always congenital. When this is recognized as an inherent deficiency inside the cartilages of the tracheal segment, it is referred to as primary or de novo. TM may be localized or generalized. [9-11] While segmental cartilage defects are extremely uncommon, one or more cartilages can be missing from the length of the trachea, causing to airway abnormalities. Secondary tracheal diseases are frequently encountered in conjunction with vascular or thoracic cage abnormalities in general. [3] Reduced or irregular connective tissues in the trachea, notably the cartilaginous rings, is also one of the main the underlying causes of TM [8].

Tracheobronchomalacia is another condition in which the major bronchi are also damaged (TBM). When excessive collapsibility is limited to one or both mainstem bronchi and/or their divisions at the lobar or segmental level, the term bronchomalacia (BM) is employed. Isolated BM, as well as extrathoracic or cervical TM, are extremely uncommon [9, 11-14].

The main two causes of TM are:

- **Congenital defects:** Congenital tracheomalacia is caused by immaturity of the tracheobronchial cartilage, which can be caused by a variety of illnesses. Systemic diseases such as Ehlers Danlos syndrome and bronchopulmonary dysplasia can cause a compromised trachea [2] In patients with corrected Oesophageal Atresia and Tracheoesophageal Fistula, respiratory symptoms are prevalent, and tracheomalacia, which can lead to life-threatening cyanotic episodes, has been described as the most common severe issue following OA and TOF repair [15, 16]. It's possible that the time between OA healing and the onset of respiratory symptoms is less than 30 days [16]. there is a high prevalence of congenital cardiac disorders that coexist. A right aortic arch, a left-sided origin of the (right) innominate artery, a right-sided origin of the left common carotid artery, or an abnormal origin of the left pulmonary artery from the right pulmonary artery, among other disorders, might cause tracheal compression [2].
- **Acquired conditions:** Inflammatory disorders including tracheobronchitis and recurrent polychondritis, as well as chronic lung diseases like bronchiectasis, can damage the structure of the airway, putting it at risk of collapsing. [2] Diffuse tracheobronchial collapse is a condition that develops due to (COPD). Gastroesophageal reflux [1]. Other factors that might cause airway stricture and tracheomalacia include extended intubation, surgery, chest trauma, foreign substances, and neoplastic diseases. [2,17- 19]

1.2 Epidemiology

Congenital tracheomalacia is uncommon, and no definitive incidence figures exist. The incidence

of tracheomalacia was predicted to be at least 1 in 2100 in all age groups [15]. (incidence of approximately 1 in 1500 to 2500 children in a study in The Netherlands) [20] tracheomalacia is more frequent in babies and children with Cystic Fibrosis (CF) than in the general population, and it's linked to airway obstruction and Pseudomonas infection, the incident of CF itself is about 1 in 3,000 [21].

TM/TBM is a common respiratory condition in children with esophageal atresia, whether they have a tracheal-esophageal fistula or not. Other airway and lung diseases, including as laryngomalacia, laryngeal clefts, bronchopulmonary dysplasia, or cystic fibrosis, may be linked to it [22-28]. Patients with tracheoesophageal fistula may have a tracheal abnormality in up to 75% of cases [2].

With increased intrathoracic pressure, such as during expiration, coughing, or sobbing, the weaker trachea is more likely to collapse. External pressures such as food boluses make it vulnerable as well. [29] The incidence of morbidity and mortality is exceptionally low. Occasionally, tracheomalacia generates enough blockage to require surgery. The tracheal cartilage normalizes, the airway enlarges, and symptoms disappear in most children by the age of three (in many before age 1 y) [1].

1.3 Diagnosis

Patients with TM often have non-specific respiratory symptoms, such as loud breathing and a barking cough, respiratory distress episodes, acute life-threatening events, and recurring and/or long-term respiratory infections. [22] Although computed tomography angiography (CTA) can be used to diagnose airway problems, administering sedation or anesthesia for CTA in this situation increases the chance of an airway catastrophe [30]. The most common symptoms of tracheomalacia are dyspnea (at rest or with exertion), cough, sputum retention, wheezing or stridor or both, recurrent pulmonary infection, bronchitis, and cyanotic spells [1,16].

Bronchoscopically or radiologically, the degree of TM/TBM can be determined. There is also no commonly agreed severity classification. Anatomical changes are classified as mild (50–75 percent reduction), moderate (75–90 percent reduction), or severe (>90 percent reduction) in clinical practice, based on subjective visual evaluation at bronchoscopy [9].

If TM is found to be intrathoracic, a computed tomographic angiography can be used to assess the vasculature's connection to the afflicted location. A dynamic computed tomography (DCT) scan can assist localize the collapsed portion of the trachea. The aorta and pulmonary arteries' connections and vascular integrity are assessed using pre- and postoperative echocardiography. Surgical intervention is required if the workup reveals the aorta laying across the afflicted trachea [29].

For adults suspected with TM Expiratory CT scans are the diagnostic test of choice [31]. For children Although history, physical examination, and supportive radiographic findings can all be used to suggest tracheomalacia, flexible fiberoptic bronchoscopy remains the "gold standard" for diagnosis. [32].

1.4 Management

Most infants with tracheomalacia do not require treatment because it normally goes away on its own by the age of 1–2 years. Recurrent respiratory infections in severe cases of tracheomalacia necessitate medical intervention, such as chest physiotherapy, long-term intubation, or tracheostomy [16]. Symptomatic medicinal therapy, positive pressure ventilation, Pharmacotherapy with cholinergic medications, [32] tracheal stents, which have had varying findings, and aortopexy are all options for treating TM. Aortopexy is now the best option for the most severe patients and the most commonly used [33]. It can be performed using an open or thoracoscopic procedure from a right, left, or anterior approach [29]. Alternative choices include endoluminal or extraluminal stenting, and tracheostomy [34].

The initial strategies focused on tracheomalacia. Direct surgical correction of vascular anomalies include division of the ring/ductus, aortic uncrossing [26].

The child's past surgical history and imaging will help determine whether to use an open or thoracoscopic technique, as well as where to enter the chest. Although thoracoscopic treatments were originally conducted on the left side, some surgeons prefer to work on the right side to avoid difficulties from working near the pulmonary artery [29]. If we put the adults also into consideration Bronchoscopy and stent implantation result in great improvement, however they come with a high risk of

complications. Lateropexia surgery, tracheal excision surgery, and surgical external stabilization surgery are all alternatives [33].

1.5 Aortopexy

Short segment TM due to congenital TOF is the most common indication for anterior aortopexy. A minor left anterior thoracotomy, a partial upper sternotomy, or thoracoscopy may be used as an approach for it [9,36,37] it was first described for the treatment of vascular tracheal compression. The ascending aorta was raised and the adventitia was sutured to the sternum's undersurface. Aortopexy reduces overall expiratory effort by easing the related element of tracheal collapse with a following decrease in total airway resistance [35]. The most typical approach to aortopexy is from the front. The ascending aorta is fixed anteriorly and can be done through a left anterior thoracotomy, partial sternotomy, or thoracoscopically [2].

1.6 Stent

In several units, airway stenting is receiving increasing attention and experience. While traditional airway stenting has historically used metal stents, which almost always result in difficulties later in life, bio-degradable stents are now being used [3]. Stenting can be used mainly in two methods:

- **External stent method:** External stabilization with polytetrafluoroethylene (PTFE) was described by Hagl. The stent is implanted outside of the trachea, and sutures are utilised to bring the tracheal adventitia closer to the stent. and thus keeping the malacic segment's lumen open. And also By using bigger grafts, this approach anticipates the issue of growth. [35]
- **Internal stenting method:** The goal of stenting is to maintain the airway until it regains structural integrity as a result of growth. In 1986, Wallace et al. developed the first expandable wire intravascular stent for use in the tracheobronchial tree. [38] Starting from coiled steel springs and advancing. [35,39] In 1997, Farga and Filler published an evidence-based study on the effects and practicality of using a metal stent in the airway. [40] There are main four problems with internal stent: They may get clogged by inspissated secretions and granulation tissue on a

regular basis, putting them at risk for secondary infections and bleeding. Another issue is stent migration and stent removal [35,41].

Stent Types:

- The Palmaz stent is a valuable tool for treating life-threatening tracheomalacia as a last resort in this tough patient population; nonetheless, their usage might lead to future hemorrhage and mortality, especially in patients with tracheotomies, therefore it must be carefully examined. [42]
- The Ultraflex nitinol stent: this stent can memorize a helical shape when heated to a higher temperature. When cooled again, it returns to its former shape. The austenitic property is critical because the nitinol stent does not increase its pressure on the airway wall following expansion in contrast with The expandable metallic stent, which on the other hand, exerts continual elastic pressure on the airway. [35] this stent can alleviate dyspnea in patients, resulting in enhanced quality of life and few problems. Even in the subglottic region, and it's also safe to use. The stent is also appropriate in certain challenging conditions, such as narrow stenosis, hourglass stenosis, curved stenosis, bilateral mainstem bronchial stenoses, and lengthy stenosis, due to its exceptional flexibility and biocompatibility. [43]

2. CONCLUSION

There's no doubt that Tracheomalacia is one of serious cases that we face in modern medicine. The first challenge in Tracheomalacia comes with the diagnosis as its being miss-diagnosed with many other respiratory diseases that might show the same symptoms. With many diagnostic techniques the bronchoscopy remains the gold standard for diagnostic along with the other radiological methods.

Different invasive treatments remains the most convenient solution in many cases with Aortopexy the most commonly used method, however recently with the emerging of biodegradable and other advanced materials for stents, different stents are emerging as good alternative solution that also being used in the most severe cases and hopefully more suitable

materials will be developed with less complications and fewer risks.

CONSENT

Not applicable.

ETHICAL APPROVAL

Not applicable.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

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