

Tracheobronchomalacia treatment: how far have we come?

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Tracheobronchomalacia (TBM) is a rather underdiagnosed endobronchial disease which treatment depends on the severity of symptoms as a consequence of severe dynamical tracheal occlusion. To this date there is no consensus regarding the therapeutic approach, hence, new state-of-the-art techniques should be taken into consideration.

Definitions

The spectrum of central airway expiratory dynamic obstruction includes TBM and excessive or hyperdynamic airway collapse. Certain degree of an airway compression characterized by an invagination of the posterior membrane and narrowing of the airway cross sectional area represents a normal physiologic process. When this process is exaggerated due to airway diseases, obesity or even in healthy individuals, the proposed term is excessive dynamic airway collapse (EDAC) (1). TBM signifies diffuse or segmental weakness of the trachea and/or main bronchi (2). While the reduction of longitudinal smooth muscle fibers tightening to prevent excessive expiratory collapse seems to be the reason for EDAC, the loss of cartilaginous structures is the main finding in TBM (3).

Epidemiology

There is scarce evidence on the prevalence of TBM or EDAC and studies in general populations are still required. For example, the incidence of TBM and HDAC was found to range from 4% to 23% in patients with various respiratory symptoms undergoing bronchoscopy (2). One of

the major reasons why this prevalence varies from one study to the other is that the symptoms are commonly mistaken and attributed to other airway diseases such as asthma or chronic obstructive pulmonary disease, and accurate diagnosis could be delayed for years (3).

Etiology

In general, TBM and EDCA can be congenital or acquired. In adults, they are usually acquired or secondary to various conditions divided in airway inflammation (irritant inhalation/aspiration, recurrent airway infections, airway diseases, collagen vascular diseases, prolonged intubation, tracheostomy) or mechanical causes (airway injury or manipulation, chronic external compression) (2-6). Speaking of airway infections, pulmonary tuberculosis may affect any part of the tracheobronchial tree and its incidence has been reported to range from 6% to 50% of cases (7). Stenosis as well as malacia has been previously described, though the clinical conditions underlying tuberculous airway stenosis often involve both cicatricial stenosis and malacia (8).

Classification

A myriad of systems based on features such as location, distribution or severity have been proposed. Although these classifications are purely academic and so far have not demonstrated to be useful in terms of guiding a specific therapeutic attitude, the most recent and increasingly applied classification aims to integrate all these conditions including functional status (World Health Organization 1-4);

extent (normal, focal, multifocal, and diffuse); morphology (EDAC and TBM crescent, saber sheath, and circumferential); cause (idiopathic or secondary); severity [normal (0–50%), mild (50–75%), moderate (75–100%), severe (100%) complete collapse] (9).

Symptoms

Due to the wide range of characteristics taking place in TBM and EDAC, the symptoms may differ from being the patient completely asymptomatic to major signs and symptoms including dyspnea, intractable cough (often barking), recurrent pulmonary infections (e.g., bronchitis, pneumonias) and difficulty expectorating sputum due to impaired mucociliary clearance, wheezing/stridor, hemoptysis and syncope due to cough (2). As stated, there are no specific symptoms orientating to TBM or EDCA, though clinical suspicion and awareness are paramount to detect them.

Diagnosis

TBM and EDAC are diagnosed by means of paired inspiratory-dynamic expiratory chest computed tomography or dynamic (aka functional) bronchoscopy (10-13). These methods are complementary and have high inter and intraobserver agreement. It is important to not over-diagnose these airway complications, although distinguishing them as a primary cause or incidental finding associated with dyspnea is often difficult (14).

Treatment

Treatment of the underlying disease is fundamental to prevent TBM and EDCA progression. However, in cases of severe malacia more active measures are needed.

Positive airway pressure

Considering the intraluminal pressure achieved, increasing positive airway pressure [either continuous positive airway pressure (CPAP) or non-invasive positive pressure ventilation (NIPPV)] may overcome the dynamic expiratory obstruction in patients with severe TBM or EDCA as a pneumatic stent, and improve both quality of life and pulmonary function tests. The use of CPAP for TBM was first described in the early 80's and showed that addition of intermittent positive pressure to routine medical therapy

may be of benefit to patients with severe TBM unresponsive to conventional medical management (15,16). NIPPV can be offered with nocturnal and intermittent application during the day, with pressure settings determined during bronchoscopic titration (17). Nowadays, in cases of refractory TBM it is considered as a temporary treatment before surgery.

Stenting

It has been stated that the preferred treatment once a severe TBM has taken place, is airway stenting (14). Although, other groups, based on the current understanding of airflow physiology, recommend that stent insertion should be reserved for patients with severe TBM not responding to the treatment of the underlying disorder and positive airway pressure (1). Similar to CPAP or NIPPV, it is considered a transient measure before surgery or definitive for patients who are not surgical candidates. There is no consensus addressing which type of stent should be used for TBM. However, so far the evidence is in favor of silicone stents rather than self-expandable metallic ones (1,2) in spite of their considerable number of complications, where infections, obstructions due to mucus plugs, migrations, and formation of granulation tissue have been described (18,19). On the other hand, expandable metallic stents are associated with numerous potential problems once inserted. Furthermore, once deployed in the airway they are difficult, if not impossible, to remove. Therefore, their usefulness in the management of patients with benign large airway disease should be carefully questioned (20).

Surgery

Patients with diffuse TBM are selected for surgical treatment if the severity of malacia exceeds that of small airway disease. As stated before, temporary tracheal stenting may help to assess benefits in individual patients, but is discouraged as long-term therapy because of the complications of endotracheal stenting. One of the surgical therapies is membranous tracheoplasty restoring the convex cartilage horseshoe shape by reefing and supporting the membranous wall with a polypropylene mesh (5). In cases of severe localized TBM, resection of the malacic segment with end-to-end anastomosis should be considered. Finally, aortopexy, a surgical procedure that consists in lifting anteriorly the aorta and suturing it to the posterior surface of the sternum has been performed especially in children

with congenital abnormalities (21), but no evidence of its suitability has been determined in adults.

New techniques

Options to surpass the complications in the follow-up period are being developed, especially for airway stenting. Yet there is no sufficient evidence to support the use of these new devices such as biodegradable or drug-eluting stents, it seems feasible in humans giving the case series reported (22). Also, bioprinting technology with 3-D prosthetics has drawn more and more attention as a fabrication methodology for producing scaffolds, cells, tissues and organs, having the advantage of precise control, repeatability and individual design (23).

Conclusions

TBM treatment can be cumbersome and no generalities shall be applied considering the multiple circumstances participating in the disease progression, other than treating the underlying condition. That is why every case should be individualized so physicians, concurring with their patients, chose the best option available. New state-of-the-art techniques are being developed with promising results, for instance 3-D bioprinting, although we still have a road ahead of us in terms of determining their true utility for this complex airway affection.

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Footnote

Conflicts of Interest: The authors have no conflicts of interest to declare.

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