TRACHEOMALACIA IN CHILDREN AND ADULTS - NOT SO RARE AS EXPECTED

Roxana-Maria Nemes¹, Paraschiva Postolache²*, Doina-Clementina Cojocaru², Mimi-Floarea Nitu³
1. Institute of Pulmonology “Marius Nasta”, Bucharest
2. University of Medicine and Pharmacy “Grigore T. Popa” - Iasi
   Faculty of Medicine
3. University of Medicine and Pharmacy, Craiova
   *Corresponding author. E-mail: postpar04@yahoo.com

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(Abstract): Tracheomalacia remains a special entity present also in children and adults. Tracheomalacia refers to a weakness of the trachea. Bronchoscopy is the “golden standard” for diagnosis. Differential diagnosis includes foreign body aspiration, difficult controlled asthma and other diseases. This disease may be congenital or it may be acquired. Acquired tracheomalacia can be treated. The main symptoms in tracheomalacia are: dyspnea, sputum production, hemoptysis and cough in adults and expiratory stridor and cough in children. Tracheomalacia could be progressive in some patients. We want to bring to your attention the tools for diagnosis and different methods of treatment. Tracheomalacia is not a rare disease and therefore we need to consider it. Keywords: TRACHEOMALACIA, STRIDOR, BRONCHOSCOPY

Tracheomalacia (TM) is still an under-diagnosed disease in both pediatric and adult patients but is not a rare disease. The term malacia is derived from the Greek world “malakia” which means soft.

In TM we can find a reduction and / or an atrophy of the longitudinal elastic fibers of the pars membranacea or impaired cartilage integrity. This cause is the reason of the dynamic narrowing of the transverse or sagittal diameters of the tracheobronchial lumen.

CLASSIFICATION
The most common classification for TM is primary (congenital disease) and secondary (acquired disease). TM is the most common congenital disease of the trachea in premature infants. Acquired TM is more frequently than congenital TM.

The respiratory distress syndrome in premature infants is the most common cause for acquired TM. Other etiology for acquired TM could be: prolonged intubation, tracheostomy, double aortic arch, left atrial hypertrophy, posttraumatic. Compression (vascular, tumors, infections, posttraumatic) could affect the tracheal wall with an increase in the intrathoracic pressure (1, 2).

DIAGNOSIS
Bronchoscopy remains the ‘golden standard’ for the diagnosis of TM (1). The
pulmonary function test (spirometry) is important because it describes the flow limitation on the expiratory part of flow volume loop with a decreased FEV1, low peak expiratory flow, reduced mid expiratory / mid inspiratory ratio and preservation of the shape of the inspiratory part (2).

It is believed that a reduction by 50-70% of the tracheal lumen is a diagnostic tool for TM. Endoscopic evaluations have resulted in the identification of 3 major models of tracheal lumen narrowing: narrowing of the lateral wall of the trachea (“scabbard type”), antero-posterior narrowing of the trachea (“floppy membrane type’) and circumferential narrowing that combines the first two variants (3).

Over the years a lot of diagnosis tools were used: tracheograms, radiographs with comparative inspiratory and expiratory views, CT (computed tomography) scan, MRI (magnetic resonance imaging) scan. CT scan is noninvasive and it reveals anatomic details and the data can be reconstructed into two-dimensional and three-dimensional images (virtual bronchoscopy) (4). Dynamic CT scan images reveal the range intrathoracic tracheal diameters during forced maneuvers. MRI is important for the evaluation of the extrinsic airway abnormalities, for example vascular compression.

Differential diagnoses in children include: foreign body aspiration, difficult asthma and recurrent croup.

PATHOPHYSIOLOGY
Tracheal diameter is influenced during ventilatory movements (exhalation, inhalation), by variations of the intrathoracic pressure. During inspiration the intrathoracic pressure decreases, size of the trachea diameters get larger, and during exhalation, as intrathoracic pressure in-
crease the caliber of the trachea will decrease. These physiological changes enhance in tracheomalacia and become obviously clear in conditions that accompany increased intrathoracic pressure: forced exhalation, coughing, Valsalva maneuver (1, 2).

CLINICAL SIGNS AND SYMPTOMS
The signs and symptoms of TM may appear during the first weeks to the first months of life (5). The most frequently symptoms are expiratory stridor, wheezing and cough. Symptomatology can vary from low-grade intermittent to continuous and severe. Children with TM have an increased relative risk of lower tract illness and a delayed recovery from intercurrent illness. We can find also: cyanosis, respiratory distress, sternal and intercostal retractions, spontaneous hyperextension of the neck, prolonged expiratory phase (6). Some children may have feeding difficulties like regurgitation and dysphagia (7, 8). In severe TM cases can appear increase sputum retention, reflex apnea and stridor during tidal breathing. Sometimes trachea is compressed by the esophagus which produces arterial desaturation, respiratory obstruction and weight loss.

TREATMENT
Intervention is not necessary in some children with TM, because when the child grows the tracheal cartilage strengthens, especially in children with mild and moderate TM, by 18-24 months (9).

Surgical intervention resolves the underlying cause (10).

Asymptomatic patients do not need therapy.

Initially, even in symptomatic patients the treatment is started with conservative
measures (chest physical therapy, control of infection, control of secretions, slow feedings, air humidifier) (11).

When conservative measures failed, surgical therapy is necessary. Tracheostomy, prosthetic stenting, tracheoplasty or tracheal resection with end-to-end anastomosis is some options regarding treatment for proximal or diffuse tracheomalacia (8, 9, 10).

Intraoperative bronchoscopy with concomitant aortopexy is the option in case of distal tracheomalacia with tracheoesophageal fistula or vascular anomalies (12, 13, 14).

In adult patients we can find tracheomalacia in patients who have chronic obstructive pulmonary disease (COPD). It is necessary to treat first the COPD (8, 9, 11).

Stents (metal, silicone) can be placed for tracheomalacia and some authors reported long-term pulmonary function improvement (15, 16, 17). Metal stents are expandable, but could provide secondary reaction: granulation tissue, fracture of the stent (18, 19, 20). Silicone stents could be placed and repositioned by rigid bronchoscopy using general anesthesia but can migrate from inset (21). There are also newer hybrid biodegradable stents, whose efficiency will be evaluated. Some patients may benefit from stenting in the long-term.

CPAP (Continuous Positive Airway Pressure) is very important as treatment for severe TM. Long-term use of CPAP could affect speech and oral feeding (22, 23).

Administration of inhaled bronchodilators in patients with TM is not helpful (24).

CONCLUSIONS
Tracheomalacia is a pathologic condition which causes dynamic narrowing of tracheal lumen. With new tools for diagnosis, TM is identified increasingly often in children and adults too. Early diagnosis of TM and individually management of disease could improve respiratory symptoms and lung function parameters.

REFERENCES